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SHUNT OPERATIONS IN MORBUS CAERULEUS

Results in 161 Cases and Clinical Follow-up.

by
TAGE MÖLLER

STOCKHOLM 1962

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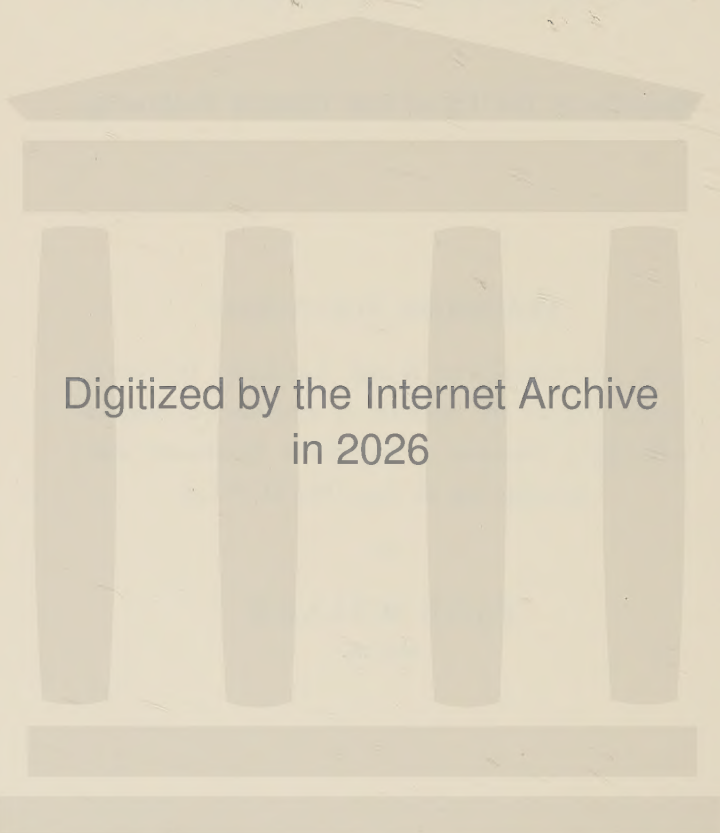
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TAGE MÖLLER

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TAGE MÖLLER

TRANSLATED FROM THE SWEDISH

BY

RAE McVEIGH



Victor Pettersons Bokindustri Aktiebolag
Stockholm 1962

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PREFACE

This investigation of the results of shunt operations in morbus caeruleus was carried out at Kronprinsessan Lovisa's Children's Hospital in Stockholm. Without close collaboration from colleagues representing the disciplines of cardiology, surgery, paediatrics and roentgenology the investigation would not have been possible. I wish to thank most warmly all of the many past and present colleagues at Kronprinsessan Lovisa's Hospital and the paediatric clinics of Karolinska, Sabbatsberg and St. Erik's Hospitals in Stockholm and of the Academic Hospital in Uppsala and Lund Lasarett who have contributed to the fulfilment of my work.

To my Chief, Professor Curt Gyllenswärd of Kronprinsessan Lovisa's Children's Hospital, I express my heartfelt thanks for support and assistance during the course of my investigation, and for kindly placing at my disposal the resources of his Clinic.

Professor Clarence Crafoord operated on 35 of the cases in the series. I am sincerely grateful to him for allowing me access to these cases.

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To Mrs. E. Reigo for her able drawing of diagrams and to Miss R. McVeigh for translation of the Swedish manuscript my sincere thanks are due.

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Tage Möller

INTRODUCTION

Ever since GROSS performed the first successful operation on the heart (August 26, 1938) by ligating a patent ductus arteriosus, interest in surgical treatment of congenital cardiac disease has steadily increased. For persons severely disabled by morbus caeruleus—"blue babies"—with stenosis or atresia of the pulmonary artery, the BLALOCK-TAUSSIG anastomosis (first performed on November 29, 1944) brought hope of considerable alleviation by diversion of incompletely oxygenated blood from the systemic to the pulmonary circulation. The first operations of this type in Sweden were performed in 1947 by P. Sandblom at Kronprinsessan Lovisa's Children's Hospital and C. Crafoord at Sabbatsberg Hospital. POTTS' (1946) modification of the shunt operation, with direct aortopulmonary anastomosis, has also been used in later years. In many "blue babies" these shunt procedures have led to improvement. Their true purpose, however, is to be regarded as palliation by increasing the flow of blood through the lungs. The anatomic defects remain even when the results otherwise are good.

More recently, shunting operations have largely been superseded by open-heart surgery in which septal defects are closed and pulmonary stenosis is corrected. This development has been made possible mainly by the evolution of cross circulation or continuous arterial reservoir perfusion (LILLEHEI ET AL. 1955), the development of safe extracorporeal techniques, the advances in intracardiac corrective surgery and the use of cardioplegia (KIRKLIN ET AL. 1959). Among the group of defects known as morbus caeruleus, the tetralogy of Fallot is widely considered to be specially suitable for intracardiac repair. Despite selection of cases the surgical mortality was high to begin with—substantially higher than in shunting procedures. In the past two years, however, the mortality from open-heart surgery has greatly declined, indicating that many problems associated with the technique have now largely been solved.

Since the tendency from all appearances is towards radical correction of the intracardiac defects, there would seem to be a need for long-term evaluation of extracardiac anastomosis in the treatment of morbus caeruleus. As well as the merits of the method, its future usefulness as compared

with intracardiac repair may thus be estimated. Some follow-up investigations of shunt operations have already been published, among the most recent of which may be mentioned those by CAMPBELL (1958), DONZELOT ET AL. (1957), PAUL, MILLER & POTTS (1961), SHUMACKER & MANDELBAUM (1960), and TAUSSIG, CRAWFORD, PELARGONIO & ZACHARIOUDAKIS (1959). Although the follow-up period in some cases was as long as 13 years, the postoperative observations frequently were less penetrating than those made preoperatively. A search of the literature has revealed few detailed reports of large clinical series examined postoperatively with heart catheterization and angiocardiography.

The present investigation comprises 161 cases of morbus caeruleus treated by Blalock-Taussig or Potts anastomosis between 1947 and 1957. The aims of follow-up were to elucidate the results of surgery and observe the postoperative condition of the surviving patients with regard to general development, physical powers, cardiovascular status and function of the anastomosis, particularly with respect to pressure and circulation in the regions involved. Because of the relative paucity of reports of post-anastomosis heart catheterization or angiocardiography, I have attempted to use these methods in as many as possible of the patients who survived the operation, and not only in those who showed the poorest results of surgery or who had to undergo a second operation.

Important among the details of the investigation was whether or not the shunt had given rise secondarily to hypertension in the pulmonary artery such as may occur in other congenital anomalies with large left-right shunt and with increased pulmonary circulation. In cases with progressive cardiac enlargement after creation of the anastomosis, the size of the shunt and the pressure in the pulmonary artery were studied as potential causal factors. Shunts created in early childhood were investigated as regards their functional adequacy in later years when demands on the circulation had increased. As the technical resources for preoperative examination were comparatively restricted in the beginning of the period from which the series was drawn, the postoperative investigations, particularly heart catheterization and selective angiocardiography (introduced by CHAVEZ, DORBECKER & CELIS 1947 and elaborated by JÖNSSON, BRODÉN & KARNELL 1949) were highly valuable as regards differential diagnosis and studies of the circulation.

On the basis of the observations made, views are advanced concerning the use of shunt operations for morbus caeruleus.

CHAPTER 1

CLINICAL MATERIAL

The series comprised 161 patients with morbus caeruleus who underwent shunt operations. In 126 cases the operations were performed at Kronprinsessan Lovisa's Children's Hospital¹ during the period April 3, 1947 to April 9, 1957. The remaining 35 cases were operated on at Sabbatsberg Hospital² or the Red Cross Nursing Home between November 5, 1947 and January 29, 1952; (for the sake of brevity I have classed these two institutions together as Sabbatsberg Hospital).

The preoperative investigations were carried out in 139 of the 161 cases at the medical department of Kronprinsessan Lovisa's Children's Hospital, in 13 cases at the paediatric department of Karolinska Hospital, in 3 cases at the medical department of Sabbatsberg Hospital, and in 2 cases at Norrtull Children's Hospital, all in Stockholm. The 4 remaining cases were investigated at the paediatric department of the Academic Hospital in Uppsala.

The nationalities of the patients were as follows: Swedish 129, Austrian 3, Belgian 7, Czechoslovakian 2, Danish 3, Finnish 7, German 1, Hungarian 3, Icelandic 1, Norwegian 2 and Polish 3. That all 32 patients from abroad were included, although satisfactory follow-up was possible in only a few of them, was because shunt operations represent an epoch in cardiac surgery. To exclude non-Swedish patients would have implied selection among the clinical material. Only as regards comparisons of height and weight between the patients and normal Swedish boys and girls were the foreign patients temporarily excluded.

The distribution of the series according to age at operation and sex is shown in figure 1. The sex ratio was 84 males and 77 females (52 and 48 per cent of the series). Statistical analysis showed no significant difference in the sex distribution. The age range was from 2 months to 16 years, except for 3 patients between 17 and 22 years. The group ≤ 9 years com-

¹ In tables, figures and text this hospital generally is denoted as *KLB*.

² Generally indicated by *S*.

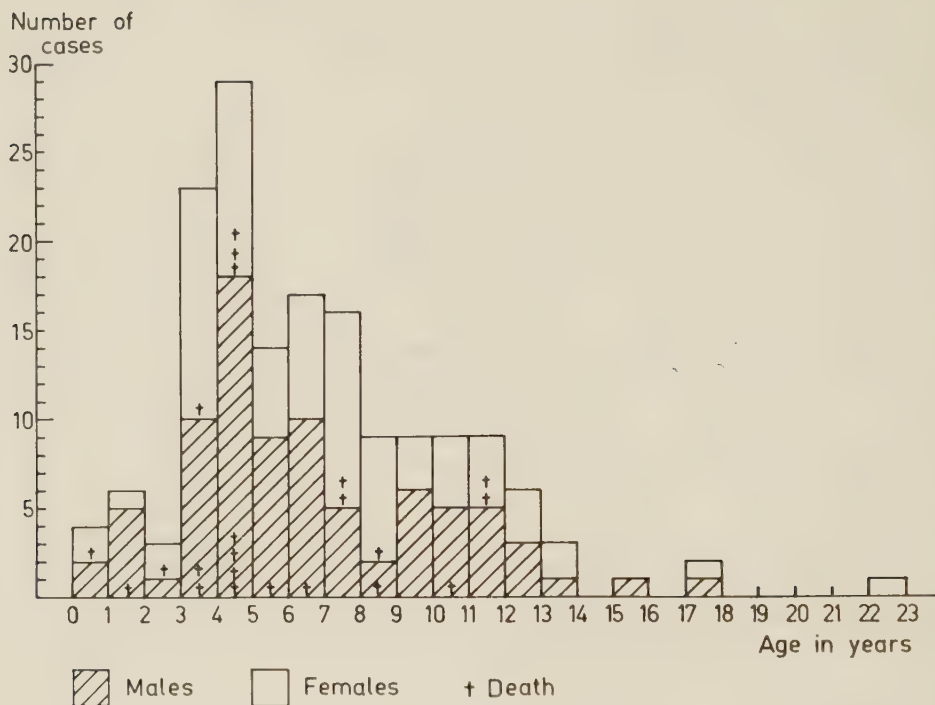


Fig. 1. Distribution of the case material according to age and sex.

prised 80.8 per cent of the total series of cases. The short life expectancy in untreated cases of morbus caeruleus doubtless was the main reason for the low age. Only exceptionally do such severely deformed persons live well into adult life. The average duration of life in the series described by ABBOTT (1936) was 12 years if the pulmonary artery was stenosed and 6 years if the pulmonary artery showed atresia. BOWIE (1961) reviewed previous reports of survival beyond the age of 40 years with "tetralogy and trilogy" of Fallot. The cases were few and illustrated the poor outlook unless these anomalies can be surgically treated. Transposition of the great vessels and tricuspid atresia carried a considerably poorer prognosis than did the tetralogy of Fallot. Fatal cerebrovascular accidents occur frequently in the tetralogy of Fallot, as was shown by ABBOTT's series. MARTELLE & LINDE (1961) stated that, in their cases of cyanotic tetralogy of Fallot, cerebrovascular accidents occurred only in the group under 5 years of age. They were correlated with hypochromic anaemia rather than with high haematocrit and increased blood viscosity.

As stated, the aim was to follow up as many as possible of the patients who survived operation. Twenty-two patients died in association with the shunt operation (tables 1, 2 and 3). Later in the observation period there were 9 deaths without relation to surgery. Attempts at re-operation because of a poor primary result or subsequent deterioration from other causes led to 7 more deaths.

TABLE 1
Type of Operation and Operative Mortality

	Blalock-Taussig		Potts		Total	
	Number of cases	Mortality	Number of cases	Mortality	Number of cases	Mortality
S	35	3 = 8.6 %	—	—	35	3 = 8.6 %
KLB	113	19 = 16.8 %	13	0	126	19 = 15.1 %
Total	148	22 = 14.9 %	13	0	161	22 = 13.7 %

TABLE 2
Number of Cases Operated on from 3/4 1947 to 9/4 1957 and Types of Surgery

Year	All cases			Blalock-Taussig			Potts		
	S	KLB	Total	S	KLB	Total	S	KLB	Total
1947	2	13 (6)	15 (6)	2	13 (6)	15 (6)			
1948	10 (1)	12 (3)	22 (4)	10 (1)	11 (3)	21 (4)		1	1
1949	15 (1)	16 (3)	31 (4)	15 (1)	16 (3)	31 (4)			
1950	7 (1)	28 (1)	35 (2)	7 (1)	27 (1)	34 (2)		1	1
1951		11 (2)	11 (2)		7 (2)	7 (2)		4	4
1952	1	13	14	1	10	11		3	3
1953		14	14		12	12		2	2
1954		10 (3)	10 (3)		10 (3)	10 (3)			
1955		7	7		5	5		2	2
1956		1	1		1	1			
1957		1 (1)	1 (1)		1 (1)	1 (1)			
Total	35 (3)	126 (19)	161 (22)	35 (3)	113 (19)	148 (22)	0	13	13

These operations refer to the initial shunt procedure. Subsequent revisions are not shown here. The figures in brackets represent the number of surgical deaths.

TABLE 3
Age at Operation and Operative Mortality

	Age in years					Total
	≤ 4	5—9	10—14	15—19	20—24	
KLB						
Number of cases ..	58 = 46.1 %	49 = 38.9 %	18 = 14.2 %	1 = 0.8 %	0	126
Deaths in age groups	12	6	1	0	0	19 = 15.1 %
S						
Number of cases ..	7 = 20.0 %	16 = 45.7 %	9 = 25.7 %	2 = 5.7 %	1 = 2.9 %	35
Deaths in age groups	1	0	2	0	0	3 = 8.6 %
Total						
Number of cases ..	65 = 40.4 %	65 = 40.4 %	27 = 16.8 %	3 = 1.8 %	1 = 0.6 %	161
Deaths in age groups	13	6	3	0	0	22 = 13.7 %

All the survivors received a written request to present themselves for re-examination.

The postoperative investigations were made by me personally in 87 cases admitted to the medical department of KLB. In 13 other cases the follow-up examination was made in the out-patient department of the same hospital, as hospitalization for various reasons was not feasible. Ten patients were hospitalized for follow-up elsewhere in Sweden. Five received out-patient examination at a Finnish and one at a Norwegian hospital. Two Swedish and 13 other patients reported their condition by letter but were unable to attend for re-examination; 3 of these foreign patients, however, sent reports of investigations made by their own doctors. Four foreign patients could not be traced, but 2 of them had previously sent written health reports to the surgeon concerned. In the cases examined at other hospitals, written records, electrocardiograms, roentgenograms, etc. were reviewed by me. Two patients from Finland and one from Germany returned to KLB for in-patient follow-up examination. The questionnaire sent to the patients who did not attend for re-examination contained enquiries concerning general health and physical capacity.

The preoperative and postoperative investigations at Kronprinsessan Lovisa's Children's Hospital, Karolinska Hospital and Uppsala Academic Hospital were carried out on largely similar principles and with the same techniques. They were therefore comparable to the extent that they were successful. The Blalock-Taussig operations performed at the first hospital

and those at Sabbatsberg were essentially similar, apart from minor technical variations, and thus were also comparable. The respective totals of this operation were 113 and 35. The remaining 13 patients underwent the Potts operation at KLB (tables 1, 2 and 3).

The 97 patients who were hospitalized for follow-up examination all underwent, in addition to routine physical examination, cardiac roentgenography, electrocardiography, tests of cardiorespiratory function, and analyses of blood and urine. No selection of cases was made for preoperative or postoperative studies; the only restrictions arose from availability of techniques or of patients. Follow-up heart catheterization was performed in 89 cases and angiocardiography in 63. These studies were made in as many cases as possible and irrespectively of whether or not they had already been done before the operation. It was thereby desired not only to investigate the postoperative circulatory status, but also to review the preoperatively made diagnosis, which in some of the early cases was found to be inaccurate or incomplete (table 4). One reason for these errors was the lower standard of technical resources at the beginning of the study period as compared with later years. The findings at follow-up examination led to a second operation in 20 cases. Because of the short observation period, the late results of these subsequent operations cannot yet be presented.

TABLE 4
*Preoperative Diagnoses Corrected after Follow-up Investigation or Autopsy*¹

Diagnosis	No. of cases	
	Preoperative	Postoperative
Tetralogy of Fallot	152	135 (19+3)
Tetralogy of Fallot + patent ductus arteriosus	1	
Probably tetralogy of Fallot	2	10 (0+2)
Pulmonic stenosis + patent foramen ovale		1
Pulmonic stenosis + atrial septal defect		1
Pulmonic stenosis + ostium atrioventriculare commune		1 (0+1)
Pulmonic stenosis + single ventricle		2 (2+0)
Tricuspid atresia or stenosis	5	8 (1+3)
Transposition of the great vessels	1	2
Obscure case with reduced pulmonary circulation		1
Total	161	161 (22+9)

¹ The first of the figures in brackets denotes primary surgical deaths and the second figure denotes late deaths.

"Probably tetralogy of Fallot" = cases in which operation was performed early in the study period, before investigation techniques were adequate for certainty in diagnosis.

The follow-up studies were concluded by the end of 1958. The observation period from the operation (1947—1957) to the latest follow-up examination varied considerably, the range being 4 months to 10 10/12 years. In the cases in which a second operation was performed, the interval between the operations was counted as the follow-up period. The mean period of observation was 5 4/12 years (median 5 10/12 years). The length of follow-up is reviewed in figure 2.

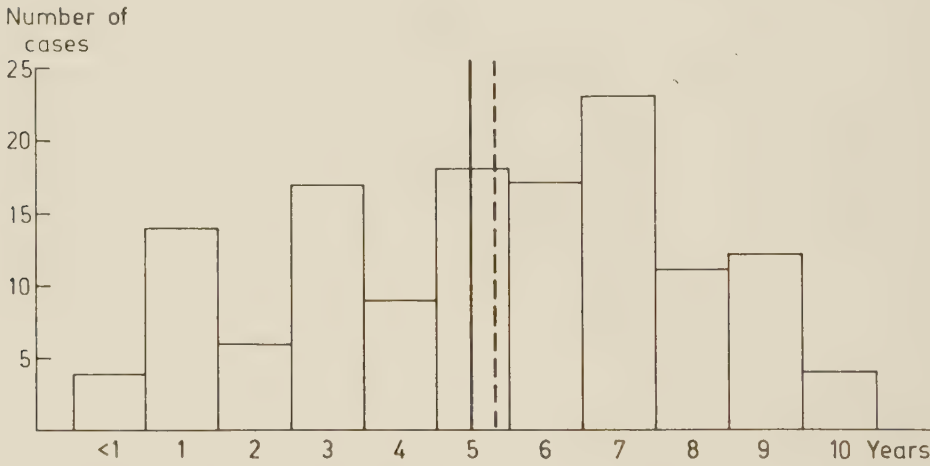


Fig. 2. Follow-up period in postoperatively examined cases.
The uninterrupted line denotes the mean period of observation and the interrupted line the median.

CHAPTER 2

DIAGNOSES IN THE CASE MATERIAL

Tetralogy of Fallot and tricuspid atresia were the main indications for shunt operations in my series of cases. Many follow-up investigations have shown that the best results of such operations are obtained in these two types of congenital heart disease. Since both the tetralogy of Fallot and tricuspid atresia may show a wide range of variations in pathologic anatomy and haemodynamics, a brief survey of the two conditions would seem to be warranted.

The Tetralogy of Fallot

The most common form of congenital cardiac disorder which gives rise to a cyanotic state is that known as the tetralogy of Fallot. Even before FALLOT in 1888 made his classic description of the condition, it had been recognized. According to KEITH (1958) it was first described by STENSEN in 1671. BENNET (1946) stated that the first clear account was given by SANDIFORT in 1777. FALLOT delineated the clinicopathologic correlation of the malformation in cases with severe obstruction of the ventricular out-flow tract. In the opinion of TAUSSIG (1947) the cases described by FALLOT represented both stenosis and atresia of the pulmonic valve. PEACOCK (1866) discussed the same conditions. WILLIUS (1948) surveyed the literature from the 18th and the early 19th century, including works by WILLIAM HUNTER (1784), FARRE (1814), and GINTRAC (1824).

In an autopsy study of 357 congenitally malformed hearts at the Mayo Clinic, FONTANA (1957) found the tetralogy of Fallot in 28 (8 per cent). BRINTON & CAMPBELL (1953) reported that at Guy's Hospital in London 25 cases of the tetralogy were found in 55 autopsies on persons with congenital cardiac disease between 1947 and 1950. This high frequency they attributed to the relatively large percentage of these cases for which medical and surgical aid is sought. OBER & MOORE (1955), in an investigation of deaths from congenital heart disease during the neonatal period, found the tetralogy of Fallot in 4 per cent. The corresponding figure in ABBOTT'S

material (1927) was 15 per cent. Persons with the tetralogy not infrequently survive longer than those with other types of cardiovascular maldevelopment (GOULD 1960).

From the clinical point of view, FALLOT found that 74 per cent of persons with malformations of morbus caeruleus type had the anatomy we now call the tetralogy of FALLOT. DONZELOT & d'ALLAINES (1954) and MANNHEIMER (1949) respectively reported corresponding frequencies of 67 and 52 per cent. GARDINER & KEITH (1951) and McLEAN & KEITH (1952) expressed the opinion that from birth to the age of 15 years the number of cases of the tetralogy equals that of all other cyanotic defects and that the tetralogy is the third most common condition in the total group.

Anatomically the tetralogy of Fallot combines obstruction of the pulmonic valve, subaortic defect in the interventricular septum, over-riding of the aorta and, as a result of these three abnormalities, hypertrophy of the right ventricle. Usually when an interventricular septal defect coexists with pulmonic valve obstruction there is free communication between the ventricles.

In earlier years the tetralogy of Fallot was considered to present a constant clinical picture. Now, however, it is recognized that persons with much the same anatomic defects can show substantial variations in the functional and clinical picture. The functional behaviour of the cardiovascular system may vary as widely in Fallot's tetralogy as in ventricular septal defect without pulmonic stenosis (McCord, VAN ELK & BLOUNT 1957). If the defect is small, the pulmonic stenosis will be the dominant anomaly. The behaviour of the cardiovascular system then is largely comparable with that in "pure" pulmonic stenosis (SCHUMACKER & LURIE 1953, McGOON & KIRKLIN 1958). When a large septal defect is present, the direction of the shunt is dependent on the degree of pulmonic stenosis. Severe stenosis associated with a septal defect, even when the aortic root is normally positioned, can give rise to a large right-left shunt and a clinical picture identical with that in Fallot's tetralogy. Because of the difficulties in determining degree of aortic over-riding, cases in which there is doubt of the aorta's position now are frequently given the designation of Fallot's tetralogy.

Pulmonic atresia is to be regarded as an extreme variant of stenosis. A common practice has been to count only cases with stenosis of the pulmonary outflow tract as Fallot's tetralogy, while classifying atresia at the valve as pseudotruncus or truncus aorticus (TAUSSIG 1947). From the functional point of view, however, the difference is of degree only. The clinical picture is determined by the state of the pulmonary circulation and is

influenced by other anomalies which may coexist with the tetralogy, such as a patent ductus arteriosus with a left-right shunt or well developed collateral bronchial arteries.

The pulmonic obstruction generally is of infundibular type (BROCK 1949, SELLORS & BELCHER 1950, BURKE, KIRKLIN & EDWARDS 1951, BERRI, CAPRILE & KREUTZER 1952, BAFFES, JOHNSON, POTTS & GIBSON 1953, BRINTON & CAMPBELL 1953, JOHNS, WILLIAMS & BLALOCK 1953). The narrowed subpulmonary outflow tract of the right ventricle has been called the "third ventricle". The calibre of the obstructed channel can show wide variations. Sometimes it is uniform throughout its course, but usually the stenosis is greatest inferiorly. In some cases there have even been observed two zones in this region with the same degree of stenosis (BRINTON & CAMPBELL). The lowest region of the subpulmonary narrowing of the outflow tract has been called the ostium of the infundibulum. The valves and the main stem of the pulmonary artery usually are of normal width but may be dilated. Instead of the usual three cusps in the pulmonic valve, most of these cases of Fallot's tetralogy show a bicuspid valve (KOLETSKY 1941).

In the less common, valvular type of stenosis, the blood flow from the right ventricle to the pulmonary artery may be greatly obstructed. In a few cases there is both infundibular and valvular stenosis. Atresia at the valve may be produced by fusion of the cusps or by failure of a communicating channel to form between the right ventricle and the pulmonary artery. In most cases of Fallot's tetralogy with valvular pulmonary stenosis the pulmonary artery is narrower than normal, but offers no true hindrance to the flow to the main branches.

Obstruction within the main stem of the pulmonary artery occurs in a few cases (CHASE 1929, EAST & BARNARD 1938, FELDMAN & SNOOK 1938, STERNBERG, POTTS, PHILIPSBORN & GIBSON 1947), and occasionally the artery is atretic, resembling a fibrous cord (GREENSPON & LEAMAN 1939, MISKALL 1945). Atresia of the lowest region of the main pulmonary artery stem was called "truncus arteriosus" by TAUSSIG (1947). COOLEY, BAINSON & HANLON (1949) used the term "pseudotruncus arteriosus". The presence of a main pulmonary artery stem, even if atretic, and of an aorta, is sufficient evidence that the truncus arteriosus has divided. In consequence, the malformation in these cases should be regarded as an extreme variant of the complex constituting the tetralogy of Fallot.

In rare cases the right or the left pulmonary artery fails to develop. BLALOCK (1948) reported that in 9 of 610 patients operated on for pulmonary stenosis one pulmonary artery was missing. When associated with

Fallot's tetralogy, this defect is most commonly seen on the left side (EMANUEL & PATTINSON 1956). The aorta is dextropositioned in 60 per cent of cases with congenital absence of the left pulmonary artery.

Constriction of the pulmonary artery trunk at its bifurcation has sometimes been seen in patients with interventricular septal defect and pulmonary stenosis (SCHUMACKER & LURIE; SØNDERGAARD 1954, COLES & WALKER 1956, WILLIAMS, LANGE & HECHT 1957). SØNDERGAARD coined the term "coarctation of the pulmonary artery" for this condition and his description was accepted by COLES & WALKER. However, I prefer to use "postvalvular stenosis of the pulmonary artery", suggested by WILLIAMS, LANGE & HECHT, since it has not been fully elucidated if the stenosis is of congenital or acquired type.

Increased thickness of the wall of the right ventricle is a conspicuous anatomic characteristic of the tetralogy of Fallot. In many cases this thickness equals or exceeds that of the left ventricular wall. The reason is that the right ventricle must pump blood at a pressure higher than normal in order to support the intra-aortic pressure. In this respect the anatomy resembles that in the normal foetus, where the right and left ventricles work together as a communicating system (PATTEN 1946).

The foramen ovale may be "valvular-competent" and patent in cases of the tetralogy of Fallot, but true atrial septal defect is an unusual complication. The combination of patent foramen ovale with the classic features of the tetralogy has sometimes been called the "pentalogy of Fallot".

In the fully typical cases of tetralogy of Fallot, the diagnosis may be made with probability bordering upon certainty solely by means of physical examination, electrocardiography and cardiac roentgenograms. Because of the variety of anomalies that are collectively termed Fallot's tetralogy, however, heart catheterization has been found valuable for elucidating the haemodynamics in individual cases. These have been well described by BING, VANDAM & GRAY (1947). The flow of blood through the pulmonary artery is more or less diminished as compared with the systemic circulation, according to the severity of the stenosis in the right ventricular outflow tract. Another factor influencing the blood flow to the lungs, however, is the augmentation in severe cases from the collateral channels (bronchial arteries). There may also be a patent ductus arteriosus. The severity of the outflow tract stenosis usually is such that the blood to a greater or lesser extent is shunted directly from the right ventricle into the over-riding aorta. The degree of aortic dextroposition and the size of the defect in the interventricular septum are also among the factors that determine the size of the right-left shunt. As a rule this exceeds one litre per minute per square

metre of body surface (KEITH 1958). At ventricular level there is also a left-right shunt, but as a rule this is small.

The pulmonary artery pressure in typical Fallot's tetralogy is normal to subnormal and the systolic pressure in the right ventricle is similar to that in the left ventricle and the aorta. The pressure gradient from the pulmonary artery to the right ventricle is distinct and abrupt in valvular pulmonary stenosis, whereas infundibular and combined valvular and infundibular stenosis produce an intermediate pressure of pulmonary artery type before the right ventricular pressure. This intermediate pressure, which was described by CONNOLLY, LEV, KIRKLIN & WOOD (1953), and others, suggests the presence of infundibular stenosis with right ventricular pressure above the stenosis. When the pulmonic stenosis is very severe, the presence of a catheter in the ostium further reduces the blood flow and the curve of pulmonary artery pressure then becomes atypical for an arterial pulse curve; the tracings are then small, rapid and irregular and show marked respiratory variations (KJELLBERG, MANNHEIMER, RUDHE & JONSSON 1955)—“pulmonic shudder” (LUISADA & LIU 1958).

Heart catheterization in Fallot's tetralogy becomes of diagnostic value first when the catheter can be passed from the right ventricle to the pulmonary artery and aorta. Because of the anatomic arrangement, however, this is frequently very difficult, especially in the children in whom the catheter must be passed from a saphenous vein. The abnormal direction of the infundibulum of the right ventricle further increases the difficulties of passing the catheter into the pulmonary artery. In severe stenosis or atresia this is practically impossible.

In order to ensure maximal accuracy in diagnosis and to chart the details of the pathologic anatomy, heart catheterization should whenever possible be supplemented by selective angiocardiography according to the principles stated by KJELLBERG, MANNHEIMER, RUDHE & JONSSON (1959).

Tricuspid Atresia

Tricuspid atresia has been regarded as one of the rarer congenital malformations, but it must be considered in the differential diagnosis of every patient with cyanotic congenital heart disease. The essential anomaly is agenesis or fusion of the tricuspid valve. Tricuspid atresia is not compatible with life unless there is also patency of the interatrial septum. In most cases an interventricular septal defect is present. The condition is further associated with hyperplasia of the mitral valve and left ventricle and underdevelopment of the right ventricle.

ABBOTT (1936) reported 16 cases of tricuspid atresia among her autopsies on patients with congenital heart disease. CAMPBELL (1953) found tricuspid atresia in 5 per cent of his series of cyanotic congenital heart disease. Corresponding statements in other clinical studies were approximately 2 per cent by SOULIÉ (1952), and 3 per cent by DONZELOT & D'ALLAINES (1954), and by KEITH, ROWE & VLAD (1958).

There is no communication between the right atrium and the right ventricle. An area of fibrosis or umbilication may constitute the boundary to the right ventricle. Sometimes the atresia is of membranous type. Minute valvular cusps may be present but are completely fused (ELSTER 1950, CHICHE 1952).

An attempt to classify the malformation was made by KÜHNE (1906), who distinguished between cases with and cases without transposition of the great vessels. EDWARDS & BURCHEL (1949) used the same grouping, but also recognized subgroups, such as cases of pulmonic atresia (without transposition), pulmonic or subpulmonic stenosis, or cases without stenosis. No transposition and subpulmonic stenosis seemed to be the most common of the numerous variants.

In tricuspid atresia the heart functions as a two-chambered organ (TAUSSIG 1936, BLOUNT ET AL. 1951, CHICHE 1952, ASTLEY, OLDHAM & PARSONS 1953, BROWN, HEATH, MORRIS & WHITAKER 1956). Since no blood can pass from the right atrium to the right ventricle, the venous blood from the right atrium enters the left atrium, most commonly through a patent foramen ovale (in 66 per cent of cases according to KEITH, ROWE & VLAD), or through a large atrial septal defect (such cases are more likely to survive for many years). Mixing of arterial and venous blood thus takes place within the heart. Difficulty in emptying causes the right atrium to become dilated and hypertrophic. The left atrium and ventricle are also enlarged. The mitral orifice is large, normally situated and competent. The right ventricle usually is underdeveloped and receives blood through the ventricular septal defect. The pulmonary circulation is reduced in most cases and as a rule is supplied by the pulmonary artery. The flow through the pulmonary artery may in turn be diminished by stenosis or by smallness of the ventricular septal defect. The pulmonary artery usually arises from the underdeveloped right ventricle, but in some cases the pulmonary artery is atretic and the circulation to the lungs is maintained by a patent ductus arteriosus or by collateral channels. The patent ductus arteriosus then is of small size and functionally inadequate, as shown by the severity of the symptoms very early in life, the absence of a continuous murmur, and the poor life expectancy (KEITH, ROWE & VLAD). The bron-

chial arteries may carry some blood to the lungs, but they are not obvious at autopsy and are of negligible functional value. Transposition of the great vessels is not of major haemodynamic importance in tricuspid atresia, since arteriovenous mixing inevitably occurs. More important prognostically is the distribution of blood between the pulmonary and the systemic circulation. The poorer the flow to the lungs, the graver becomes the prognosis. Pulmonic stenosis is common in these cases, whether the great vessels are normally situated or are transposed.

The clinical features of tricuspid atresia frequently are the same as in the tetralogy of Fallot, but cyanosis appears earlier. KEITH, ROWE & VLAD found that signs of failure were exceptional in cases with moderate or diminished pulmonary flow. The triad of central cyanosis, a diminutive right ventricle and left axis deviation in the electrocardiogram has led to more frequent clinical recognition of tricuspid atresia. A clinical diagnosis that rests mainly on the left ventricular preponderance will prove wrong in some of the cases, however, and angiocardiology is of great value in reaching a final conclusion. Heart catheterization is less helpful in diagnosis than angiocardiology, which must often be done more than once in order to clarify the diagnosis. The difficulties in diagnosis are illustrated by the not infrequent cases in which earlier decisions must be revised at follow-up examination or autopsy.

CHAPTER 3

SURGICAL TREATMENT

Introductory Notes

After experimental observations on the effects of connecting by suture the left main pulmonary artery to the systemic circulation (LEVY & BLALOCK 1937), such anastomosis was for the first time performed on a human subject with morbus caeruleus in 1944. The result was published by BLALOCK & TAUSSIG in 1945. The technique for creation of an artificial ductus arteriosus was described by the same writers in 1947. About the same time, a variant of the shunt operation was introduced in which direct anastomosis was made between the aorta and the pulmonary artery (POTTS, SMITH & GIBSON 1946). For the sake of simplicity, I use the names BLALOCK and POTTS to denote the respective operations.

The original Blalock anastomosis operation and the Potts variant of it both create an artificial shunt to augment pulmonary circulation which is insufficient because of stenosis or atresia of the pulmonary artery. These palliative operations, therefore, are designed to increase the supply of oxygenated blood to the aorta. When they are successful, the preoperative cyanosis and clubbing of fingers and toes largely disappears and tolerance for physical exercise is greatly improved. The communication from the aorta to the pulmonary artery is made by direct anastomosis in the Potts operation and indirectly, using the subclavian or another systemic artery, in the Blalock procedure. End-to-side union between the subclavian and the pulmonary artery involves little risk to the circulation in the ipsilateral arm. If the innominate artery is used, however, there is appreciable risk of impaired blood flow to the brain. Excessively wide anastomosis with a thick branch of the aorta, or a large side-to-side aorticopulmonary orifice, may entail too great an additional burden on the heart. POTTS, SMITH & GIBSON stated some guiding principles concerning the size of the anastomosis, so as to prevent overloading of the pulmonary circulation. The subclavian artery, because of its relative narrowness, implies less risk in this respect than does the innominate or the carotid artery.

Methods

Of the 161 patients in my series, 148 underwent the Blalock operation and 13 the Potts operation.

In all 13 children submitted to the Potts operation, the anastomosis was made directly between the aorta and the left pulmonary artery. The pre-operative investigations or surgical exploration had shown in these cases that the anatomic pattern was unsuitable for a Blalock anastomosis: As a rule the subclavian artery was too short or too narrow. The age at operation ranged from 3 to 11 years, the mean age being 5 years. The length of the incision into the aorta and the pulmonary artery varied between 3 and 8 mm. In the 4 cases in which 3 to 4 mm incisions were made, there was no definite postoperative improvement, and in 2 of them thrombosis was suspected to have occurred in the anastomosis. In 6 cases with incisions measuring from 5 to 8 mm, the results were very good and the anastomosis seemed to function well. In the remaining 3 cases the length of the incisions was not recorded, but the postoperative results were satisfactory. According to POTTS ET AL. (1955), the ideal incision is 6.3 mm long. This gives an anastomosis channel of 4 mm.

Blalock anastomosis was done on the right side in 69 cases and on the left side in 79 cases. The choice of side was dependent in the individual cases on the surgeon's evaluation of technical considerations. Important factors were the position of the aorta and the size and site of the subclavian artery or other branch of the aorta which was thought to be suitable for anastomosis. The appearance, size and site of the pulmonary artery and the prospects of mobilizing it for anastomosis were other determinative circumstances. In most cases the preoperative cardiologic investigations provided information in these respects. Angiocardiology then played a decisive part in the choice between left-sided or right-sided anastomosis. Frequently, however, the final decision was made after the chest had been opened and the anatomic arrangement could be directly visualized. In the Blalock operations the anastomosis generally was made on the side opposite to the aorta: Thus, of the 79 patients with left-sided anastomosis, the aorta was on the right side in 58.2 per cent, and of the 69 cases with right-sided anastomosis, 91.3 per cent had left-sided aorta.

For 137 of the 148 Blalock operations the typical end-to-side subclavian-pulmonary artery anastomosis was chosen. The surgical technique was essentially similar at Kronprinsessan Lovisa's Children's Hospital and at Sabbatsberg Hospital. Because of technical difficulties, other procedures than end-to-side subclavian-pulmonary artery anastomosis were selected

for the 11 cases of Fallot's tetralogy which are surveyed in table 5. One of these patients died on the first postoperative day, 5 were improved by the operation and 5 were unimproved; 3 of these latter underwent further surgery later. In Case 139 there was marked improvement during the first years after the operation, but this was followed by rapidly progressive

TABLE 5
Variants of Blalock Anastomosis in Fallot's Tetralogy

Name	No.	Age & Sex	Anastomosis operation	Reason for variation of technique	Result
A.S.	8	♀ 6 $\frac{5}{12}$	End-to-side right innominate artery and central portion of superior branch of right pulmonary artery.	Right subclavian artery originated high up.	Improved
A.U.	7	♂ 4 $\frac{5}{12}$	End-to-end right subclavian artery and right pulmonary artery.	Short right subclavian artery.	Improved
C.R.	144	♂ 4 $\frac{7}{12}$	End-to-end left subclavian artery and superior branch of left pulmonary artery.	Wide left subclavian and narrow, thin-walled left pulmonary artery.	Unimproved
J.Ö.	139	♂ 4 $\frac{6}{12}$	End-to-side right external carotid artery and right pulmonary artery.	Short right subclavian artery with high origin.	Transitorily improved
K.K.	68	♂ 3 $\frac{3}{12}$	End-to-side right axillary artery and right pulmonary artery.	Wide right subclavian artery.	Unimproved
K.K.Å.	64	♂ 4	End-to-side left common carotid artery and left pulmonary artery.	Atresia of left subclavian artery.	Died of pulmonary oedema 1 day after operation
L.G.	81	♀ 3 $\frac{8}{12}$	End-to-end right subclavian artery and central portion of superior branch of right pulmonary artery.	High aortic arch and short right subclavian artery.	Unimproved
M.E.	94	♀ 3 $\frac{10}{12}$	End-to-side left common carotid artery and left pulmonary artery.	Atresia of left subclavian artery.	Improved
O.J.O.	104	♂ 6 $\frac{10}{12}$	End-to-side right subclavian artery and right pulmonary artery after division of right common carotid artery.	Short right subclavian artery	Unimproved
T.R.	130	♀ 3 $\frac{1}{12}$	End-to-end right subclavian artery and superior branch of right pulmonary artery.	Short right subclavian artery.	Improved
W.J.	140	♂ 4 $\frac{6}{12}$	End-to-side right subclavian artery to superior branch of right pulmonary vein.	Right pulmonary artery not found.	Unimproved

cardiac enlargement produced by the large left-right shunt through the wide anastomosis between the external carotid and the pulmonary artery. The surgical procedure in this case was complicated by the high origin of the subclavian artery, which rendered it unsuitable for anastomosis. The wall of the pulmonary artery tore as it was being joined to the external carotid, and in the new union that was made the orifice was larger than was originally intended. Pulmonary hypertension and cardiac insufficiency gradually developed and were accompanied by frequent and severe attacks of paroxysmal tachycardia. Further surgery is considered to be contraindicated by the cardiac insufficiency. In Case 64 the subclavian artery was atresic and the common carotid was considered to be the only vessel suitable for anastomosis with the pulmonary artery. Already during the operation the anastomosis was feared to be too wide in view of the patient's relatively moderate degree of preoperative disablement. Death occurred on the following day from cardiac failure and pulmonary oedema due to overloading of the pulmonary circuit. In 4 cases end-to-end anastomosis was made with the pulmonary artery—the superior branch in 3 cases. In another operation (Case 140) the pulmonary artery could not be found and anastomosis was therefore made between the subclavian artery and the superior branch of the pulmonary vein, which opened into the superior vena cava. During the first 3 years after the operation the patient's condition was thought to be somewhat improved. Subsequently, however, there was marked deterioration. At a second operation an infundibular stenosis was resected *ad modum* Brock, but cardiac arrest occurred and the patient died on the operating table. Autopsy revealed a narrow but patent anastomosis between the right subclavian artery and the vein supplying the upper lobe of the right lung.

In 2 of the other 137 cases it was found at autopsy that the subclavian artery had been inadvertently anastomosed to the superior pulmonary vein (cf. table 6).

Four of the 148 patients treated by Blalock operation underwent a preliminary exploratory thoracotomy. In 3 patients it was then found that an aberrant subclavian artery could not be mobilized; the anastomosis was subsequently made between the subclavian and the pulmonary artery on the other side. In the fourth patient (Case 116) the reason for closing the chest without anastomosis was the observation of extremely strong pulsations in the pulmonary artery, which were suspected to be caused by a patent ductus arteriosus. At preoperative heart catheterization the pressure in the pulmonary artery could not be registered, and therefore the pulsations were thought possibly to indicate pulmonary hypertension. At a second

Name	No.	Age & Sex	Preoperative incapacity	Clinical findings	Operation
A. I.	159	♂ 3 $\frac{7}{12}$	Extreme	Typical tetralogy	Blalock end-to-side, left
E. H.	24	♂ 8 $\frac{6}{12}$	Extreme	Typical tetralogy	Blalock end-to-side, right (ligature slipped)
E. B.	25	♀ 4 $\frac{4}{12}$	Moderate	Typical tetralogy	Blalock end-to-side, left
H. J. E.	44	♂ 4 $\frac{8}{12}$	Moderate	Typical tetralogy	Blalock end-to-side, left
H. A.	47	♀ 3 $\frac{9}{12}$	Moderate	Tetralogy	Blalock end-to-side, right
J. S. G.	52	♂ 4 $\frac{3}{12}$	Extreme	Typical tetralogy	Blalock end-to-side, left
K. R.	58	♀ 4 $\frac{2}{12}$	Extreme	Typical tetralogy	Blalock end-to-side, right
K. I.	60	♀ 7 $\frac{4}{12}$	Extreme	Typical tetralogy	Blalock end-to-end, left
K. K. Å.	64	♂ 5	Moderate	Typical tetralogy	Blalock end-to-side, left
K. G.	66	♀ 11 $\frac{1}{12}$	Extreme	Typical tetralogy	Blalock end-to-side, right
K. I. L.	152	♀ $\frac{6}{12}$	Extreme	Tricuspid atresia	Blalock end-to-side, left
K. P.	70	♂ 3 $\frac{3}{12}$	Extreme	Typical tetralogy	Blalock end-to-side, left
L. B.	83	♂ 6	Moderate	Typical tetralogy	Blalock end-to-side, right
L. M. H.	86	♂ 5 $\frac{2}{12}$	Extreme	Tetralogy with pulmo- nary atresia	Blalock end-to-side, right
M. A.	90	♀ 2 $\frac{4}{12}$	Extreme	Typical tetralogy	Blalock end-to-side, right
N. O.	95	♂ 3 $\frac{5}{12}$	Extreme	Typical tetralogy	Blalock end-to-side, right
P. M.	111	♀ 4 $\frac{2}{12}$	Moderate	Tetralogy plus patent ductus arteriosus	Blalock end-to-end, left
R. I. B.	116	♀ 9 $\frac{7}{12}$	Moderate	Typical tetralogy	Blalock end-to-side, left
S. L.	120	♀ 7 $\frac{10}{12}$	Moderate	Typical tetralogy	Blalock end-to-side, left
S. U.	121	♀ 12 $\frac{4}{12}$	Extreme	Tetralogy	Blalock end-to-side, left
S. R.	156	♂ 1 $\frac{2}{12}$	Extreme	Typical tetralogy	Blalock end-to-side, left
V. T.	135	♂ 4 $\frac{6}{12}$	Moderate	Typical tetralogy	Blalock end-to-side, left

with Operation

Course	Autopsy findings
Circulatory and respiratory collapse during operation. Died 3 hours later.	Tetralogy. Right pneumothorax, left haemothorax. Total atelectasis of right lung and partial atelectasis of left lung.
Thrombosis at site of anastomosis. Cerebral damage. Died of circulatory insufficiency 12 hours after operation.	Tetralogy. Thrombosis in anastomosis. Atelectasis and broncho-pneumonia.
Acute deterioration on 2nd postoperative day. Died of circulatory insufficiency.	Tetralogy. Subclavian artery anastomosed to superior pulmonary vein. Intimal rupture in subclavian artery.
Respiratory distress and pyrexia during 1st postoperative day. Suddenly worse on 4th day—air hunger and circulatory failure.	Tetralogy. Atelectasis of left lung. Acute bilateral bronchitis and broncheolitis.
Died 2 hours after operation with signs of haemothorax.	Tetralogy. Right haemothorax and atelectasis.
Temperature rising to hyperpyrexia and shock. Died on 2nd postoperative day.	Tetralogy. Bilateral atelectasis.
Progressive cardiac enlargement and insufficiency. Died on 2nd postoperative day.	Tetralogy. Pulmonary oedema.
Anastomosis attempt failed. Left pulmonary artery ligated and divided. Died during operation.	Tetralogy with pulmonary atresia.
Died of pulmonary oedema on 1st postoperative day.	Tetralogy. Atelectasis of left lung and pulmonary oedema.
Evacuation of blood from thorax 8 days after operation. Died of shock.	Cor triloculare biventriculorum and valvular pulmonic stenosis. Right haemothorax and atelectasis. Thrombosis in anastomosis.
Respiratory insufficiency. Died on 1st postoperative day.	Tricuspid atresia. Interatrial and interventricular septal defect. Hepatic and splenic stasis. Atelectasis of left lung.
Cardiac arrest after completed anastomosis. Ventricular fibrillation during operation.	Cor triloculare biatriorum, anomalous pulmonary veins, verrucous endocarditis of right ventricular outflow tract.
Thrombosis in anastomosis after 1 day. Died of pulmonary oedema on 3rd postoperative day.	Tetralogy. Right haemothorax and atelectasis. Oedema of left lung.
Died of shock immediately after operation.	Tetralogy with pulmonary atresia. Left haemothorax.
Died of pulmonary oedema a few hours after operation.	Tetralogy. Encephalomalacia.
Died of circulatory and respiratory insufficiency a few hours after operation.	Tetralogy. Right haemothorax.
Died as thorax was being closed.	Tetralogy with pulmonary atresia. Subclavian anastomosis to central portion of pulmonary artery, which terminated blindly.
Ventricular fibrillation and death during operation.	Tetralogy.
Died of pulmonary artery thrombosis and circulatory insufficiency on 1st postoperative day.	Tetralogy. Thrombosis of left pulmonary artery.
Died 2 days after operation. Hyperpyrexia. Left pneumothorax.	Tetralogy. Bronchitis and signs of cardiac failure.
Signs of grave cerebral injury—unconsciousness and convulsions. Died on 6th postoperative day.	Tetralogy. Left pleural empyema. Severe organic and cerebral stasis.
Died 2 hours after operation.	Tetralogy. Subclavian artery anastomosed to superior pulmonary vein.

catheterization 4 years later, the pressure in the pulmonary artery was found to be normal to subnormal, thus confirming the earlier diagnosis of Fallot's tetralogy. A second attempt at subclavian-pulmonary artery anastomosis on the same side as before, however, terminated for no clear reason in fatal cardiac arrest.

Nonfatal Complications of Surgery

In table 7 are shown the 27 cases in which the records described non-fatal surgical complications. Horner's syndrome (sinking in of the eyeball, ptosis of the upper eyelid, slight elevation of the lower lid, constriction of the pupil, narrowing of the palpebral fissure, and anhidrosis caused by paralysis of the cervical sympathetic) was the most common of these complications (14 cases), and may safely be said to have occurred, unrecorded, in milder form in yet more cases. Irritation or damage to the cervical sympathetic during operation must be presumed to have been the causal mechanism in these cases. Only very occasionally was deformity from Horner's syndrome permanent. Respiratory complications in the form of atelectasis and haemothorax were second in order of incidence (8 cases); when haemorrhage was severe, blood was evacuated from the chest by suction. One patient had hemiplegia, probably due to cerebral embolism, and another showed clear evidence of pulmonary embolism. Clinical signs of pericarditis followed slight trauma to the pericardium during operation in one patient, but at follow-up examination no lasting effects were seen.

TABLE 7
Nonfatal Surgical Complications

Name	No.	Age & Sex	Diagnosis	Operation	Complications
A.K.	1	♀ 2 ⁹ / ₁₂	Fallot's tetralogy	Blalock end-to-side, left	Laryngeal stridor. Tracheotomy 1st postoperative day.
A.I.	8	♀ 6 ⁵ / ₁₂	Fallot's tetralogy	Blalock end-to-side, right	Atelectasis of right upper lobe.
A.H.	6	12 ⁶ / ₁₂	Fallot's tetralogy	Blalock end-to-side, right	Horner's syndrome, right.
A.U.	7	♂ 4 ⁴ / ₁₂	Fallot's tetralogy	Blalock end-to-end, right	Horner's syndrome, right.
A.J.O.	9	♂ 5 ⁶ / ₁₂	Fallot's tetralogy	Blalock end-to-side, right	Suspected acute pericarditis.

Name	No.	Age & Sex	Diagnosis	Operation	Complications
B. B.	17	♀ 7 $\frac{1}{2}$	Fallot's tetralogy	Blalock end-to-side, right	Horner's syndrome, right.
B. P.	14	♂ 4 $\frac{3}{2}$	Fallot's tetralogy	Blalock end-to-side, right	Horner's syndrome, right.
C. T.	20	♂ 5 $\frac{3}{2}$	Fallot's tetralogy	Blalock end-to-side, left	Horner's syndrome, left. Pneumothorax.
C. E.	150	♀ $\frac{1}{2}$	Tricuspid atresia	Blalock end-to-side, left	Atelectasis of right middle lobe.
E. G.	145	11 $\frac{3}{2}$ ♀	Fallot's tetralogy	Blalock end-to-side, left	Horner's syndrome, left.
E. B.	28	♀ 8 $\frac{8}{2}$	Fallot's tetralogy	Blalock end-to-side, right	Horner's syndrome, left.
E. N.	161	♀ 3 $\frac{8}{2}$	Fallot's tetralogy	Blalock end-to-side, left	Horner's syndrome, left.
E. Y.	31	♂ 5 $\frac{1}{2}$	Fallot's tetralogy	Blalock end-to-side, left	Haemothorax, left.
H. J.	39	♂ 6 $\frac{1}{2}$	Fallot's tetralogy	Blalock end-to-side, left	Cerebral embolism with right hemiplegia.
H. B.	42	♀ 7 $\frac{2}{2}$	Fallot's tetralogy	Blalock end-to-side, right	Haemothorax and pneumothorax, right.
J. H.	50	♂ 7 $\frac{2}{2}$	Fallot's tetralogy	Blalock end-to-side, right	Laryngeal stridor. Tracheotomy 2nd postoperative day.
J. B.	57	♀ 8	Fallot's tetralogy	Blalock end-to-side, left	Atelectasis of right upper lobe. Right hemiplegia.
K. G.	59	♀ 5 $\frac{5}{2}$	Fallot's tetralogy	Blalock end-to-side, left	Haemothorax, left.
L. U. B.	76	♀ 3 $\frac{6}{2}$	Fallot's tetralogy	Potts, left	Paralytic ileus, anuria.
L. K. G.	80	♂ 4 $\frac{5}{2}$	Fallot's tetralogy	Blalock end-to-side, left	Horner's syndrome, left.
L. G.	81	♀ 3 $\frac{8}{2}$	Fallot's tetralogy	Blalock end-to-end, right	Pulmonary embolism.
M. J.	88	♂ 4 $\frac{1}{2}$	Fallot's tetralogy	Blalock end-to-side, left	Horner's syndrome, left.
M. M.	154	♀ 7 $\frac{5}{2}$	Fallot's tetralogy	Blalock end-to-side, left	Horner's syndrome, left.
N. G.	98	13 $\frac{8}{2}$ ♂	Fallot's tetralogy	Blalock end-to-side, right	Horner's syndrome, right.
N. B.	99	♂ 6 $\frac{8}{2}$	Fallot's tetralogy	Blalock end-to-side, right	Horner's syndrome, right.
P. L.	112	♂ 4	Fallot's tetralogy	Blalock end-to-side, left	Haemothorax, left. Anastomosis resutured.
S. M.	129	♀ 4 $\frac{3}{2}$	Fallot's tetralogy	Blalock end-to-side, right	Horner's syndrome, right.

Surgical Mortality

The primary surgical mortality was 13.7 per cent. The sex distribution was 10 males and 12 females, and there was no significant sex preponderance among the fatal cases. Nor was there any statistically significant difference between the *KLB* and the *S* cases as regards primary mortality. For comparisons of age in these cases, the series was divided into 4 groups. Here, too, significant differences failed to emerge. Some tendency towards higher surgical mortality seemed to be discernible, however, in the patients younger than 5 years as compared with the other age groups.

From tables 1, 2, 3 and 6, which show the deaths during shunt operations or in the immediate postoperative period, the causes of death may be summarized as follows.

Causes of Death in 22 Cases

	<i>Main causes</i>	<i>Contributory causes</i>
	(no. of patients)	
Cerebral complications		3
Cardiac arrhythmia during operation	2	
Cardiac failure with or without pulmonary oedema	4	1
Haemorrhage	4	2
Thrombosis of the anastomosis	1	2
Respiratory complications	7	5
Shock		3
Miscellaneous and unexplained	4	

To distinguish between main and contributory causes of death in the complicated picture presented by such patients may be extremely difficult. In the present series, however, *respiratory complications* (atelectasis and infection) seemed to predominate. Prophylactic administration of penicillin thus failed to prevent infection in some cases. In one case pleural empyema developed but its diagnosis was delayed by the coexistence of cerebral damage with unconsciousness and convulsions. *Haemothorax* was the main cause of death in 4 cases and a contributory cause in 2. It usually arose from a leaking anastomosis; in Case 66 an unsuccessful attempt was made to evacuate the blood on the eighth postoperative day.

When the Blalock operation was first described, it was known from work by EPPINGER, BURWELL & GROSS (1941) that a patent ductus arteriosus can increase the work of the heart by as much as 400 per cent. It was

hoped, however, that cardiac enlargement following the creation of a Blalock anastomosis would be transitory, and that *cardiac failure* would not be a common complication. To judge from subsequent reports, this hope seems to have been largely justified. In one of the cases with cardiac failure and pulmonary oedema in the present series, the common carotid artery was used for anastomosis and the resultant left-right shunt was excessive (Case 64). This risk was also pointed out by BAHNSON & ZIEGLER (1950), who wrote that when the heart is preoperatively enlarged a large shunt through, for instance, the carotid or the innominate artery can give rise to the mentioned complications. The same writers further stressed the risks from a large artificial shunt in extreme pulmonic stenosis. These circumstances occurred in 2 patients of the present series.

Thrombosis in the anastomosis gave rise in 1 patient to progressive blocking of the left main branch of the pulmonary artery, which led to circulatory failure and death on the first postoperative day. In 2 other cases thrombosis at the site of anastomosis was a contributory cause of death. Technical difficulties and unsuitable position of the anastomosis could cause kinking or pulling of the vessels with resultant impairment of the circulation and thereby a tendency to thrombus formation. By auscultation over the site of anastomosis it was usually possible to decide when thrombosis had occurred, since the continuous murmur then disappeared. Deterioration of the patient's general condition, particularly as regards cyanosis, could be concurrently seen. Thrombosis at the site of anastomosis thus was a serious complication.

Cerebral complications were not found to be the main cause of death in any case, but were contributory causes in 3 cases. This, of course, is to be regarded more as an assumption than as a conclusion, since cerebral complications often are very difficult to demonstrate at autopsy. The relative infrequency of such complications seems surprising in view of the tendency to cerebral damage in untreated cases of morbus caeruleus in which arterial oxygen saturation is low and there is appreciable polycythaemia. Both of these conditions predispose the brain to damage from cardiorespiratory and circulatory disturbances during surgery. In some of the patients who preoperatively had attacks of cyanosis associated with unconsciousness and fits, it is likely that cerebral damage was already present when surgery was done. In the series operated on by Blalock and reported by BAHNSON & ZIEGLER, cerebral complications were fatal in 4.4 per cent of the patients and were one of the main causes of death.

Cardiac arrhythmia was relatively unimportant as a cause of death, since ventricular fibrillation occurred in only 2 patients. Preoperative digi-

talization was carried out only in a minority of the patients. Such preparation did not seem to be significant for the outcome of surgery, except in the decompensated patients.

The group called *miscellaneous and unexplained* was heterogeneous. In 2 of the 4 cases the subclavian artery was anastomosed by mistake to the pulmonary vein, and in 1 of these 2 autopsy showed rupture of the intima at the site of subclavian anastomosis. Technical difficulties arose in the third and fourth cases. At operation Fallot's tetralogy with pulmonary atresia was found. One of the patients died during an attempt at end-to-end anastomosis of the subclavian artery with the pulmonary artery. In the other patient the subclavian artery was joined end-to-end with the central portion of the pulmonary artery which, however, later was found to terminate blindly.

Summary

1. Of 161 patients with morbus caeruleus, 148 were operated on according to Blalock and 13 according to Potts. The operative mortality was 13.7 per cent (22 patients). No deaths occurred in association with the Potts operation. The surgical mortality in the children younger than 5 years seemed to be somewhat higher than in the other age groups. There was no statistically significant difference as regards sex in the fatal cases.
2. In 11 cases the usual technique of Blalock anastomosis was varied because of technical difficulties. Of these patients 5 were improved by surgery, 5 were unimproved and 1 died.
3. In 4 cases the Blalock operation was preceded by exploratory thoracotomy. The reason why anastomosis was not initially done was technical anatomic difficulties in 3 cases, and in the fourth case it was suspected that patent ductus arteriosus had caused pulmonary arterial hypertension.
4. Nonfatal complications followed surgery in 27 cases. After Horner's syndrome, respiratory complications in the form of atelectasis and haemothorax were the most common.
5. Twenty-two patients died in immediate association with the anastomosis operation. The most common cause of death was respiratory complications. Next in order of frequency was bleeding from leaking anastomosis. Cardiac failure and pulmonary oedema resulted from an excessively wide anastomosis with the carotid artery in one case.
6. Cerebral complications and cardiac arrhythmia were rare in this series.
7. Thrombosis of the anastomosis site was a serious complication, directly fatal in one case and contributing to the fatal outcome in 2 other cases.

CHAPTER 4

CLINICAL FEATURES

Antenatal History

GREGG (1942) and SWAN, MOORE, MAYO & BLACK (1943) reported an association between rubella in pregnant women and congenital malformations in the children of these pregnancies. KEITH, ROWE & VLAD (1958), however, expressed the opinion that cases of congenital cardiac anomaly following a definite history of rubella in the first trimester of the particular pregnancy were few. Cases of this type were described by BRINTON & CAMPBELL (1953) and GIBSON & LEWIS (1952). KJELLBERG, MANNHEIMER, RUDHE & JONSSON (1959) found that in 10 of 668 cases of congenital cardiac disease the mothers had contracted a virus infection early in pregnancy (rubella in nine cases and mumps in one case). LARSSON & MANNHEIMER (1949) reported that, among 80 children with morbus caeruleus, rubella had complicated the first month of pregnancy in one case and severe influenza had occurred in the third month in two other cases.

In my series of cases there were no known instances of rubella in the mother during pregnancy. Three mothers, however (of Cases 54, 146 and 164), reported influenza or severe "colds" during the second or third months of pregnancy. Among other aetiologic factors may be mentioned bleeding in the second to third month of pregnancy (Cases 13, 109 and 120) and chronic nephritis revealed towards the end of the first trimester (Case 71). RICHARDS, MERRITT, SAMUELS & LANGMANN (1955) and POLANI & CAMPBELL (1955) found increased risk of Fallot's tetralogy with late maternal age, but this was not confirmed in my investigation.

No other antenatal circumstances seemed to be noteworthy.

Delivery was assisted by forceps in six cases (19, 27, 58, 71, 116 and 164). In three other cases (12, 33 and 61) caesarean section was done because of pelvic disproportion.

Birth Weight

In the 148 cases in which the birth weight was known, it ranged from 1.57 to 4.94 kg (fig. 3). In 81.8 per cent of these children the range was

2.5 to 4.0 kg. Only 6 per cent weighed 4 kg or more. Prematurely born infants (birth weight less than 2.5 kg) constituted 12.2 per cent of the series.

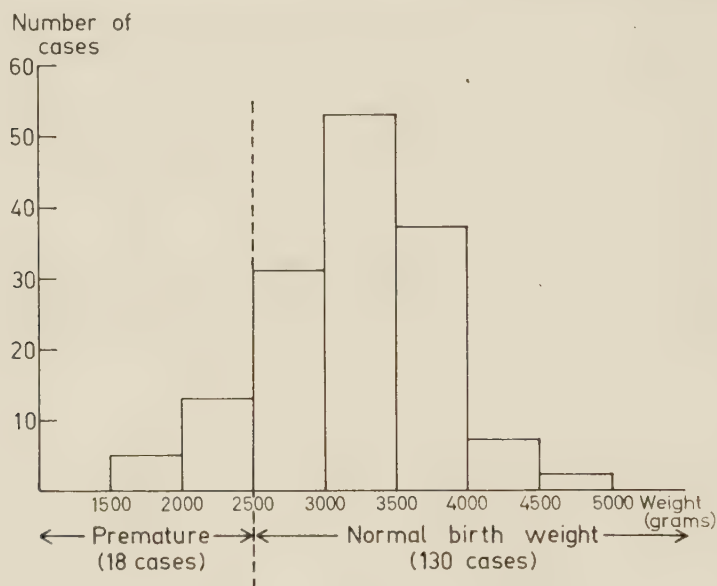


Fig. 3. Birth weight in 148 children operated on for morbus caeruleus. In 13 cases no information was available.

This figure is somewhat high in comparison with normal infants. LICHTENSTEIN (1941) found 6.2 per cent prematurity in a study of children born in Stockholm between 1936 and 1938. In similar studies from Gothenburg and the periphery of Stockholm, VON SYDOW (1940) and GYLLENSWÄRD (1946) respectively reported frequencies of 5.7 and 7.7 per cent. On the other hand, LEECH (1935) found that 13.3 per cent of children with congenital cardiac malformations were prematurely born. MANNHEIMER (1949) reported 10.9 per cent prematurity in children with morbus caeruleus, and KEITH, ROWE & VLAD stated that in about 10 per cent of children operated on for Fallot's tetralogy birth had been premature.

One of the children in my series weighed 1.60 kg at birth and had a still-born twin; the cause of still birth was extreme prematurity (weight 800 g).

Familial Occurrence

In association with the follow-up study, enquiries were made concerning cardiac disorders in the patients' siblings. The distribution of siblings is shown in figure 4. Four of my patients had had a sibling with congenital

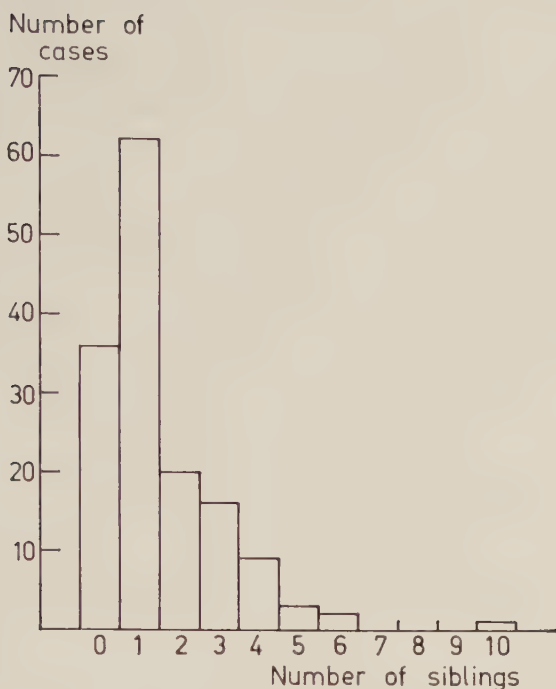


Fig. 4. Distribution of siblings in 159 children operated on for morbus caeruleus. In 2 cases no information was available.

cardiac malformation. In two cases the sibling was born before, and in two after, my patient. These siblings had died at ages ranging from 7 months to 20 years. Three of them had disease of the morbus caeruleus group without special diagnosis, and the fourth, who had verified Fallot's tetralogy, died shortly before a projected operation could be undertaken. In no case was there a surviving sibling with known congenital cardiac disease. McKEOWN, MacMAHON & PARSONS (1953) found that 1.8 per cent of siblings born after their cases of congenital cardiac disease were similarly affected. This figure they calculated to be six times the frequency in the general population. POLANI & CAMPBELL, in a similar study, found a familial incidence of 2.26 per cent, implying a sevenfold increase above the general frequency. Because of the relatively few cases in my series, I have not attempted to make comparisons of this type.

In 6 of my 161 cases the child was one of heterozygous twins. Congenital anomaly of the heart or other organs was in no case demonstrated in the other twin. UCHIDA & ROWE (1957) concluded from a detailed analysis of the literature on twins that only in a minority of cases were both twins affected.

Sex Distribution

Among the 356 infants and children with congenital heart disease who were studied by KEITH, ROWE & VLAD in Toronto, the male/female ratio was 49 and 51 per cent. This is in conformity with figures found by other writers (WHITE 1945, TAUSSIG 1947, LANDTMAN 1947). In some specified forms of congenital heart disease differing proportions have been reported (ABBOTT 1936, TAUSSIG 1947, McKEOWN 1953, ANDERSSON 1954, POLANI & CAMPBELL; KEITH, ROWE & VLAD). Male preponderance thus was found by GOULD (1960) among cases of aortic stenosis or transposition of the great vessels, by REIFENSTEIN, LEVINE & GROSS (1947) in coarctation of the aorta, and by FONTANA (1957) in the tetralogy of Fallot. Female preponderance was observed in patent ductus arteriosus by CRAFOORD, MANNHEIMER & WIKLUND (1944), EKSTRÖM (1952), GROSS (1952) and COSH (1957), and in atrial septal defect by WEIDMAN, SWAN, DUSHANE & WOOD (1957).

My series comprised 84 male and 77 female patients (52 and 48 per cent). The percentage relationship is unaltered if only the cases of Fallot's tetralogy are considered. KEITH, ROWE & VLAD likewise found no sex disparity in their series of tetralogy cases. POLANI & CAMPBELL, however, reported that the male/female ratio in their series was 59/41 per cent.

Cyanosis

The clinical picture in Fallot's tetralogy and comparable maldevelopmental states is mainly determined by the degree of pulmonic obstruction. In tricuspid stenosis or atresia the quantitative relationship between the ventricular blood flow to the lungs and the systemic circulation is similarly reflected.

The clinical features of severe or complete obstruction at the pulmonic valve have been described by many writers (BROWN 1945, GREEN 1945, HUMPHREYS 1947, TAUSSIG 1948, WOOD 1950, GIBSON 1952, WELLS 1952, CAMPBELL & DEUCHAR 1953). The main clinical manifestations, of which cyanosis is the most prominent, are due to anoxaemia, polycythaemia, and difficulty in the direction of oxygenated blood to the systemic circulation. In severe or total pulmonic valve obstruction, and in most forms of tricuspid atresia, cyanosis is manifest early in life, generally from birth. In the less severely deformed cases the cyanosis appears later; frequently it is first seen during increased physical or mental exertion, for instance when the infant begins to walk. KEITH, ROWE & VLAD found that, in more than

90 per cent of children with severe malformations of Fallot type, cyanosis appeared during the first six months of life. Deterioration with appearance of cyanosis later in the first year was attributed by TAUSSIG (1948) to concomitant closure of the ductus arteriosus. GASUL, DILLON, VRLA & HAIT (1957), however, based their explanation of this later appearance of cyanosis on the probability that the obstruction at the pulmonic valve can increase and thus cause the blood supply to the lungs to be progressively reduced as the child grows. Even if the calibre of the outflow tract remains the same, the natural increase in body growth and the consequently heavier demands on cardiac output imply some accentuation of the obstruction in relation to the flow, so that a right-left shunt can arise or become intensified.

The age at which cyanosis was first observed in my series of patients ranged from immediately after birth to more than 3 years. In 64 per cent of the series cyanosis was manifest before the age of 7 months. This figure is in good conformity with observations published by other writers.

As a rule, cyanosis was the first sign of congenital cardiac disorder. Its occurrence was fairly closely followed by the diagnosis of cardiovascular anomaly (table 8), except in infants less than one month old. Part of the reason for the delay in recognition of the significance of cyanosis in these young children presumably was that the first check at infant welfare centres seldom was made before the age of one month. Later in the first half year the diagnosis was more frequently made, and then at routine examinations or when medical advice was sought specifically because of cyanosis.

Cyanosis during rest was observed before operation in all but four of the patients. Relatively moderate physical exertion, however, produced cyano-

TABLE 8

Time of Appearance of Cyanosis and of Medical Diagnosis of Congenital Heart Disease

Age	Cyanosis noticed	Diagnosis made
Birth—1 month	64	49
1—7 months	39	60
7—12 „	14	15
1—2 years	13	17
2—3 „	18	10
>3 „	12	9
No record	1	1
Total	161	161

sis in all cases. In the mildest cases the cyanosis was confined to the lips and nails and in the most severely affected it appeared as a purplish discolouration over the whole body, with pronounced mucosal involvement. I have recognized four degrees of cyanosis when the patients were as far as possible in a basal state, i.e., resting and conscious. The preoperative and postoperative cyanosis is presented in table 9. It is seen that after operation there was a marked shift towards mild cyanosis or acyanosis. These two groups together comprised only 18 cases before operation but 99 afterwards. The corresponding figures for the patients with moderate or severe cyanosis were 143 and 35.

Statistical analysis was made of the postoperative improvement with respect to degree of cyanosis in relation to the age at operation (table 9). Significant difference was found between the age group ≤ 4 years and the age group > 8 years. This showed that operation in children aged 4 years or less gave poorer results as regards cyanosis than did operation after the age of 8 years. Between the other age groups the analysis showed no significant differences.

The intensity of the cyanosis varied with the amount of available haemoglobin and the percentage of reduced haemoglobin in the circulating blood. Concerning the relationship between haemoglobin and oxygen saturation in the systemic circulation, the reader is referred to page 76. According to TAUSSIG (1959), "the rate at which polycythaemia develops depends upon the degree of oxygen unsaturation of the arterial blood and the demands of the body for oxygen. The red blood cell count, the amount of available haemoglobin, and the haematocrit reading usually remain within normal limits for the first six months of life. It is for this reason that cyanosis is so much less conspicuous in infants than in children. Indeed, only in patients with severe pulmonary stenosis is there marked polycythaemia by two years of age. In young children and adults the development of polycythaemia is the rule. The amount of available haemoglobin and the haematocrit reading are proportionally increased".

Dyspnoea

In dyspnoea produced by physical exertion, the distinction between a pathologic and a normal response is not always easily made. Dyspnoea occurring during rest frequently accompanies congenital cardiac disease in infants and may be very severe if cardiac insufficiency is present. Paroxysms of tachypnoea, beginning and ending suddenly, are rare in most types of cardiac insufficiency, but are very common in "blue babies"

without insufficiency. Dyspnoea is a prominent symptom in morbus caeruleus; to begin with it arises when the infant is fed or cries, and later it occurs during physical exertion such as attempts at walking. During early infancy paroxysmal dyspnoea associated with pronounced cyanosis is most common in the severely deformed cases. According to TAUSSIG (1948) these symptoms tend to undergo spontaneous improvement between the ages of 18 months and 2 years, which may be attributed to the development of collateral channels.

In the present series of patients with right-left shunt and diminished pulmonary circulation, the peripheral oxygen saturation was lower than normal and dyspnoea at rest on the whole showed parallelism with cyanosis. The dyspnoea did not necessarily indicate the existence of cardiac insufficiency. An investigation of this type, of course, provides only a rough estimate of the incidence of dyspnoea in the resting state before and after operation. Precise calculations of respiratory frequency were not made and, indeed, were not feasible. Dyspnoea was considered to be present when breathing rates were clearly excessive for the child's age and general physical development. The patients who had mild cyanosis as a rule had no dyspnoea during rest. Of the 143 patients with moderately severe or severe cyanosis preoperatively, 54 had varying degrees of dyspnoea during rest, and in 4 cases there were no records concerning dyspnoea. Postoperatively only 12 of the entire series were found to have dyspnoea during rest. In 8 cases there was no postoperative information on this point. The other 116 surviving patients had no postoperative dyspnoea in the resting state.

Clubbing of Fingers and Toes

Most patients who have congenital cardiac disease and have been cyanotic for some length of time have clubbing of the fingers and toes. Absence of clubbing suggests that the cyanosis is of recent occurrence. Clubbing designates proliferation of capillaries and soft tissues, dilatation and thickening of the vascular walls and, in advanced cases, hypertrophy of the periosteum around the distal phalanges of fingers and toes. The gingival mucosa is also hyperplastic in these cases. Although clubbing is most commonly associated with the cyanotic group of congenital heart diseases, it may also be seen in other disorders (e.g., of the lungs or, rarely, the stomach). Beaking of the nails is, as a rule, a precursory phase of clubbing.

Among my patients there was only insignificant difference before operation in the frequency of clubbing of the fingers and toes as compared with beaking of the nails (140 and 143 cases, respectively). After operation,

however, 66 patients showed clubbing and 91 beaking, the reason being that, of the component changes of clubbing, nail beaking is the last to undergo regression.

Physical Development

Physical development is generally considered to be retarded in morbus caeruleus. Delayed increase in height has even been stated to be one of the most characteristic features of the group. ABBOTT (1936) found that 27 of 110 patients (24.5 per cent) with various types of congenital pulmonary stenosis were physically retarded. LANDTMAN (1947) reported analogous observations in 101 of 121 cases (86 per cent). LUND (1952), using the Wetzel grid for comparison, concluded that physique was never optimal and growth was slower than in the general population. From his series of cases in Toronto, KEITH (1958) concluded that "infants with the tetralogy usually gain weight slowly, over half of our postmortem group and about one third of our clinical group behaving in this manner".

The results of surgery with respect to height and weight in the present series are illustrated by figures 5 and 6. Here the values are superimposed upon charts showing height and weight in healthy Swedish boys and girls (BROMAN, DAHLBERG & LICHTENSTEIN 1942, KARLBERG & PERMAN 1959). To remove one possible source of error, patients of other nationalities were excluded. So that both preoperative and postoperative values could be obtainable, the fatal cases were likewise excluded.

The height and weight before operation were below the normal limit of variation (2σ) in some cases. As a rule the values lay between the mean and the lower variation limit. Postoperatively there seemed to be no essential change in these respects. As the observation period ranged from 1 to 10 years after operation, the conditions for comparison were not uniform, but it may be said that shunt operations for morbus caeruleus did not seem to have an important influence on physical development as regards height and weight. Similar observations were made by LUND.

Squatting

TAUSSIG (1947) described squatting as "a common habit among cyanotic children who suffer from inadequate circulation to the lungs. All these children are able to get their breath more easily in this position". Squatting thus is seen in certain types of cyanotic congenital heart disease, particularly tetralogy of Fallot.

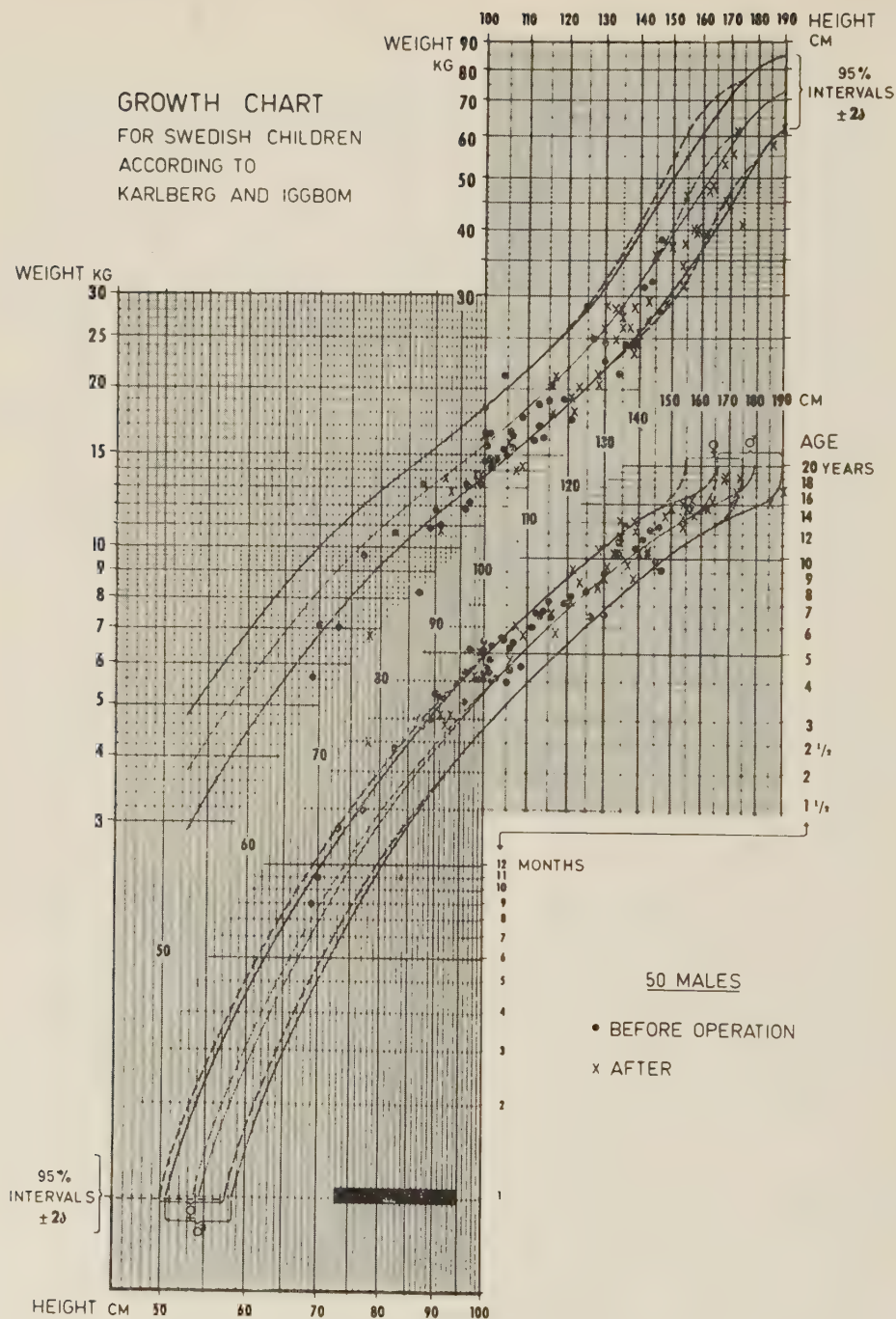


Fig. 5. Physical development in 50 males before and after operation for morbus caeruleus. Comparisons with findings by Karlberg & Iggbom in healthy children.

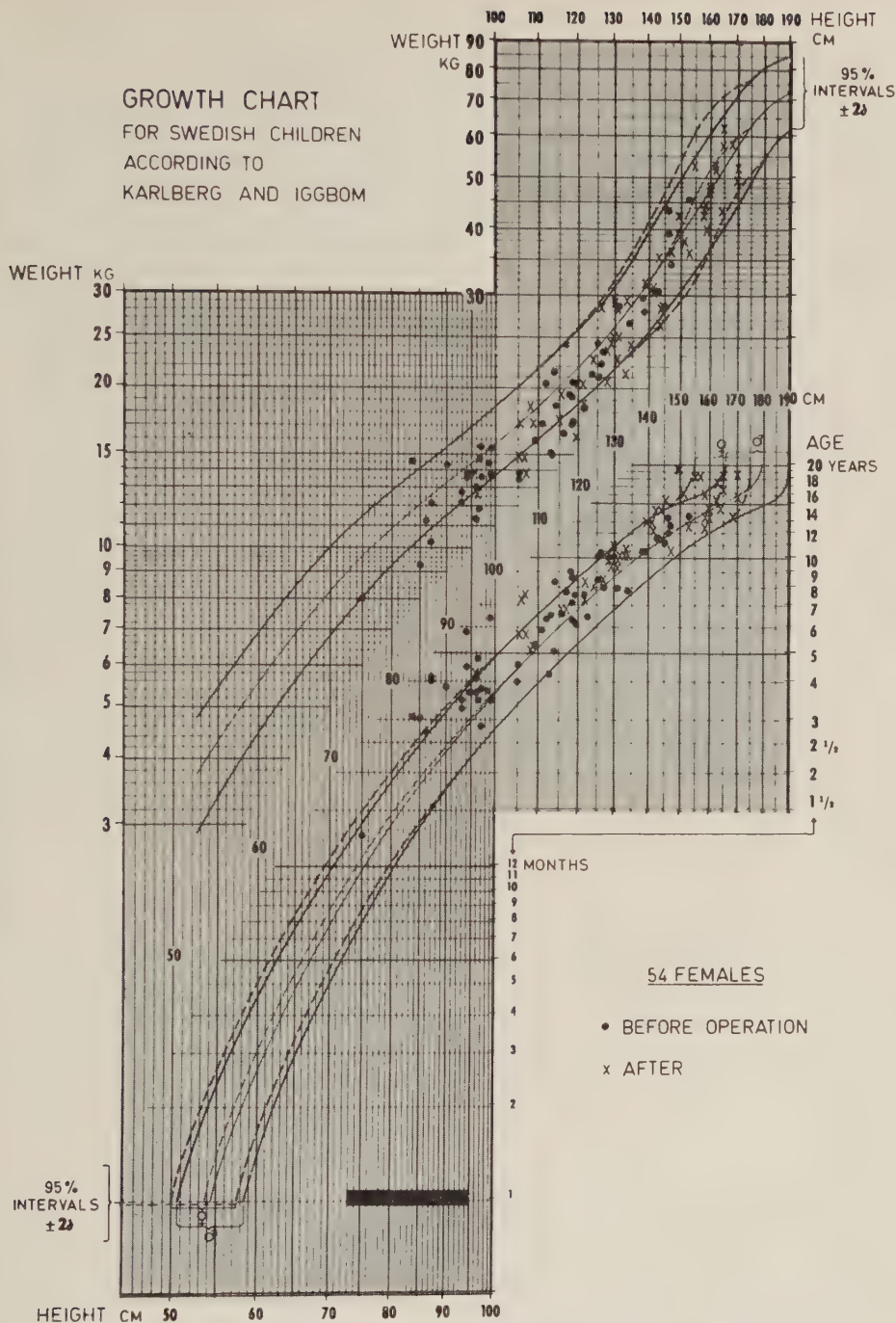


Fig. 6. Physical development in 54 females before and after operation for morbus caeruleus. Comparisons with findings by Karlberg & Iggbom in healthy children.

MANNHEIMER found squatting in 81.6 per cent of his cases of tetralogy of Fallot. MÉTIANU & DURAND reported a corresponding frequency of 70 to 80 per cent and stated that squatting was commonest in the ages 4 to 12 years. CHICHE (1952) and MÉTIANU, DURAND, COLLADO-MADERA & GUILLEMOT (1953) observed that children with tricuspid atresia frequently squatted. KEITH, ROWE & VLAD, on the other hand, stated that only 1 of 8 patients in the walking age squatted.

NADAS (1957) wrote that squatting first makes its appearance at about 18 months or when the child starts walking. Social pressure usually, though not invariably, abolishes it at 8 to 10 years of age.

BURCHELL (1950) observed that the oxygen saturation in the systemic arteries increased during squatting. According to LURIE (1953) the venous return to the heart is facilitated in the squatting position. BROTMACHER (1957) found an increase in systemic blood flow confined to the upper part of the body and combined with an increase in oxygen tension. This he attributed either to increased systemic venous return or, possibly, to augmentation of the peripheral resistance with consequent diminution of the degree of right-left shunt.

In my series squatting was observed in the following frequency.

	<i>Before operation</i>	<i>After operation</i>
Squatter	123	23
Non-squatter	25	107
Not known	3	5
Non-ambulant	10	1
Dead	—	25
Total	161	161

In 108 cases with habitual squatting before operation, postoperative comparison could be made in this respect. In figure 7 preoperative and postoperative squatting is correlated to the walking capacity. Postoperative squatting was observed in 21 cases with varying degrees of exercise tolerance.

Exercise Tolerance

In children with reduced pulmonary circulation, the tolerance for physical exercise varies according to the degree of pulmonic stenosis and the state of the collateral circulation. The tolerance may also vary from day to day in the same individual. Heat, cold and dampness tend to have unfavourable effects in this respect. In many children with tetralogy of Fallot the arterial oxygen saturation falls so rapidly during physical

TABLE 10

Walking Capacity in Different Age Groups

Walking capacity (meters)	Age (years)	Non-ambulant	Not known	0-50			50-100			100-500			500-1000			1000-5000			5000-10000			>10000		
				0-4			4-8			8-12			>12			0-4			4-8			8-12		
				Deaths			24			1			2			3			4			5		
Non-ambulant	0-4	1	1	5	1	1	1	1	1	1	1	1	1	1	1	1	1	1	1	1	1	1	1	1
Not known	0-4		1																					
	4-8	1																						
0	0-4	18		2																				
—	4-8	30		6																				
50	8-12	15		1																				
	>12	2		1																				
50	0-4	1		1																				
—	4-8	18		2																				
100	8-12	8																						
	>12	4																						
100	0-4	6																						
—	4-8	13		2																				
500	8-12	6		3																				
	>12	3																						
500	4-8	2																						
—	8-12	6																						
1000	>12	2																						
1000	4-8	9		1																				
—	8-12	3																						
5000	8-12																							

The vertical column "Number of cases" = preoperative.

The horizontal columns show postoperative cases.

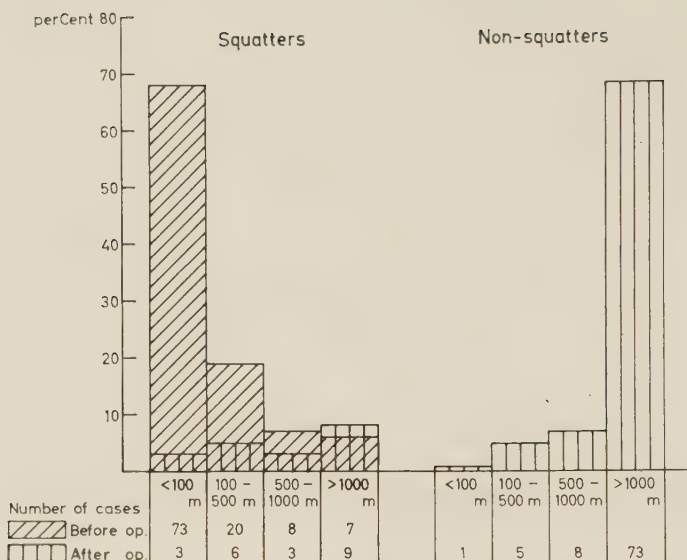


Fig. 7. 108 cases of preoperative squatting correlated to walking capacity (in metres) before and after operation.

exercise that they are abruptly compelled to rest. For these reasons it is exceedingly difficult, if not impossible, to use standardized tests of physical capacity in such cases.

I have therefore tried to assess the children's exercise tolerance by careful questioning of patients and relatives concerning walking capacity before and after operation. The questioning was not confined to one follow-up occasion, but was made in association with each personal or postal contact. By walking capacity is meant the maximum distance which the individual can accomplish at his own walking pace without pausing to rest or squat. Table 10 shows the walking capacity in different age groups before and after operation. The appreciable postoperative improvement was particularly evident in the patients who could walk no more than 100 metres before operation. In some cases the postoperative tolerance for physical exercise was so good as to be practically comparable with that in persons free from cardiac disorder. Not a few of the patients at the time of follow-up had jobs which under normal circumstances required good physical capacity. The arterial oxygen saturation in the systemic circulation combined with the degree of cyanosis constitutes another good indicator of the physical capacity. The higher the arterial oxygen saturation and the less severe the cyanosis, the better as a rule is the tolerance for physical exercise.

CHAPTER 5

CARDIAC FINDINGS

Thrills

Thrills and murmurs are basically the same phenomenon recognized in different ways. In many cases a murmur is heard although no thrill can be felt, but where thrill is present a murmur is always audible, usually strongly so.

Murmurs were heard in almost all of the patients in this series. Thrills were felt only in cases with loud systolic murmur. They were then palpable over the second to fourth left intercostal space along the sternal margin, except in the cases with dextrocardia, when thrill was felt to the right of the sternum. According to the case records, 79 patients had preoperative systolic thrill (slight in 11 cases and distinct in 68). In 76 cases no thrill was felt, and in 6 the presence or absence of thrill was not noted.

At follow-up examination after surgery, thrill arising from a functioning anastomosis was rarely encountered. On the other hand, thrill as a manifestation of loud stenosal murmur from the pulmonic region was felt in 58 cases (slight in 11, distinct in 47). In 60 cases no postoperative thrill was felt, and in 18 (chiefly patients from foreign countries) there were no records concerning postoperative thrill. The remaining 25 patients were dead at the time of follow-up.

Of the 58 cases with postoperative thrill, 15 had not had this sign before operation, and of the 60 cases without postoperative thrill 20 had had thrill preoperatively. Whether these discrepancies were attributable to difference of examiners before and after operation, or were in accordance with the facts, could not be determined from the available data. In the cases in which thrill was first felt postoperatively the age range at operation was 3 to 12 years (mean 6.8 years), and in those in which a preoperative thrill disappeared after surgery the corresponding ages were 3 to 13 years (mean 7.7 years). The age factor thus seemed to be of minor importance in this respect. According to GASUL, DILLON, VRLA & HAIT (1957), infundibular pulmonic stenosis may arise and progress in cases of ventricular septal

defect. Reduced pulmonary circulation due to the right-left shunt may result in diminution of a pulmonic stenosal murmur.

Cardiovascular Sounds

The first heart sound in cases of the tetralogy of Fallot has been described as normal and most distinct over the apex (MÉTIANU & DURAND 1954, REINHOLD & NADAS 1954, KJELLBERG, MANNHEIMER, RUDHE & JONSSON 1959). Sometimes, because of the fact that the aorta is carrying almost all, or in fact all, of the cardiac outflow, a murmur of high flow is generated in the aorta. An early systolic click (ejection sound) originating in the aorta may occur for a similar reason (LUISADA & MAGRI 1952). The ascending aorta lies nearer to the sternum than under normal conditions, which favours diffusion of sounds arising in the aorta. PATTINSON & EMANUEL (1957) considered it probable that the enlargement of the ascending aorta is a factor of importance for the occurrence of systolic click.

The second heart sound in Fallot's tetralogy has been described in different ways. According to MANNHEIMER (1949), BROWN (1950) and REINHOLD & NADAS (1954) it is usually diminished over the pulmonic region or, in some cases, quite inaudible. Other writers have described the second heart sound as distinct in many cases, and frequently loud or accentuated (TAUSSIG 1947, BAKER, BROCK & CAMPBELL 1949, WOOD 1950, REINHOLD 1958). According to KEITH, ROWE & VLAD (1958) the sound may be weak, but as a rule it is loud and only exceptionally is it split. Phonocardiography shows that the single second sound arises from closure of the aortic valves (LEATHAM 1954).

In pulmonic stenosis with intact ventricular septum, by contrast, the second heart sound is often split. This is caused by prolongation of right ventricular systole due to the heightened resistance against ejection of the stroke volume (MANNHEIMER & JONSSON 1954). The degree of splitting of the second sound is the same in infundibular and valvular stenosis. Correlation between the degree of stenosis and the width of duplication of the second sound over the pulmonic region has been demonstrated by simultaneous phonocardiography and measurements of right ventricular pressure (LEATHAM & WEITSMAN 1957).

In tricuspid atresia the heart sounds are not characteristically abnormal, except that the second sound is usually single (aorta) and the pulmonic component is totally or partially lacking (cf. pulmonic atresia) and is never accentuated.

Murmurs

The systolic murmur in the tetralogy of Fallot probably arises mainly or wholly at the stenotic pulmonary outflow tract. Because of the aortic overriding there is presumably little obstruction to outflow through the aorta, and consequently a murmur is not likely to originate at the ventricular septal defect (McKUSIK 1958).

VOGELPOEL & SCHRIRE (1960) found certain auscultatory and phonocardiographic characteristics in the systolic murmur of Fallot's tetralogy. They compared the severity of the tetralogy, as determined by clinical methods, with the length and loudness of the murmur and other auscultatory signs. Increasingly severe stenosis was found to be associated with a shorter, earlier and softer systolic murmur. In extreme tetralogy the murmur was invariably soft and very short, being confined to early systole and ending in midsystole. In severe tetralogy the murmur was usually loud and short, reaching its peak before midsystole and terminating before the aortic second sound. In tetralogy of moderate severity the murmur was loud and longer, with crescendo at midsystole and termination before or at the aortic second sound. In mild cases the murmur was loud and more prolonged, loudest in the latter half of systole and extending into, but rarely much beyond, the aortic component.

In pulmonic stenosis with intact ventricular septum, VOGELPOEL & SCHRIRE found that the severity of the stenosis, as determined by the right ventricular pressure, was correlated to the duration of right ventricular systole as determined both by the length of the systolic murmur and the width of splitting of the second heart sound. Increase in the severity of stenosis led to progressive lengthening of the murmur in relation to the aortic second sound and delay in pulmonic valve closure. In mild stenosis the murmur was soft to loud, reaching a crescendo in midsystole and ending before or at the aortic component of the second sound. In moderately severe stenosis the murmur was louder, with greater crescendo and duration, and extended a short way beyond the aortic component, which it never obscured. In severe stenosis the murmur was loud and there was great delay in crescendo and duration, which resulted in a kite-shaped rather than a diamond-shaped configuration. The murmur invariably extended well beyond the aortic component, which it partially or completely obscured, and always ended before a very soft pulmonic component, which was widely separated from the aortic component. In very severe stenosis the murmur was loud and so prolonged that it invariably obscured the aortic component, but ended before a very delayed diminutive pulmonary component, which might or might not be heard.

In tricuspid atresia there may even be no murmur, but as a rule a systolic murmur is heard, often fairly loud and without typical location. The organic murmurs seem frequently to have maximal intensity in the third and fourth left interspaces at the left sternal margin (KEITH, ROWE & VLAD). Both a ventricular septal defect and pulmonic stenosis may be responsible for the murmur (KJELLBERG, MANNHEIMER, RUDHE & JONSSON).

In the present study a distinct systolic murmur was found preoperatively by auscultation in 157 of the 161 cases, and in most of them it was phonocardiographically verified. The murmur was heard in the second to fourth left interspaces, except in the dextrocardiac Cases 113 and 115, in which it was most distinct to the right of the sternum. The records of Case 60 stated that only a diastolic murmur was heard, but this was not confirmed by phonocardiography. The patient died during operation and autopsy revealed pulmonic atresia with well-developed bronchial artery collaterals. In another patient who died at operation (Case 111) the same autopsy findings of pulmonic atresia with well-developed bronchial arteries were recorded: These collaterals could in all probability explain the continuous murmur which was present preoperatively, with maximum over the base of the heart. In 2 patients (Cases 86 and 126) the heart sounds were practically clear on auscultation and phonocardiography. The first of these patients died immediately after operation and the autopsy findings were pulmonic atresia with ventricular septal defect and over-riding of the aorta. In Case 126 the investigations showed typical Fallot's tetralogy with severe valvular and infundibular pulmonic stenosis and a large right-left shunt.

Duplication of the first heart sound (systolic click, aortic ejection sound) was heard preoperatively in 8 of the 156 cases examined by phonocardiography (5.1 per cent). The ejection sound occurred between 0.06 and 0.1 second after the first heart sound. In 6 of these 8 cases roentgenographic and angiocardiographic studies showed distinct widening of the aorta, which lay far forward against the sternum, a circumstance which thus seemed to favour the occurrence of ejection sound.

Accentuation of the heart sounds otherwise seems to be uncommon in morbus caeruleus. According to the records of the 161 cases in the present series, accentuation of the second sound over the pulmonic region was heard preoperatively in only 24 cases. Comparative phonocardiographic calculations were not made, since most of the phonocardiograms were recorded only for study of murmurs.

Duplication of the second heart sound, however, was found in only one case of Fallot's tetralogy (Case 75), with 0.06 second between the two components. In Case 106, which was originally diagnosed as Fallot's tetra-

logy, but in which surgical exploration revealed pulmonic stenosis with patent foramen ovale and right-left shunt, the phonocardiogram showed no duplication of the second heart sound and no typical site of the systolic murmur as in severe pulmonic stenosis.

Systolic murmurs in the tetralogy of Fallot can, according to VOGELPOEL & SCHRIRE, be classified from phonocardiographic tracings as having maximum amplitude in early, mid or late systole. They found this distribution to correspond to pulmonic stenosis of severe, moderate and mild degree, respectively. When the Fallot cases in the present series were distributed according to this classification of murmurs, it was found that the mean value for preoperative oxygen saturation in the femoral artery or descending aorta was 69 per cent in the cases with murmur early in systole, 72.1 per cent in those with midsystolic murmur, and 73.5 per cent in the cases with late systolic murmur. This difference in oxygen saturation is significant and agrees well with the findings of VOGELPOEL & SCHRIRE.

Shortly after the anastomosis operation, 134 patients had an organic systolic murmur. In 1 case the heart sounds were clear, without distinct murmur (Fig. 8), and in another case no records were available. The other

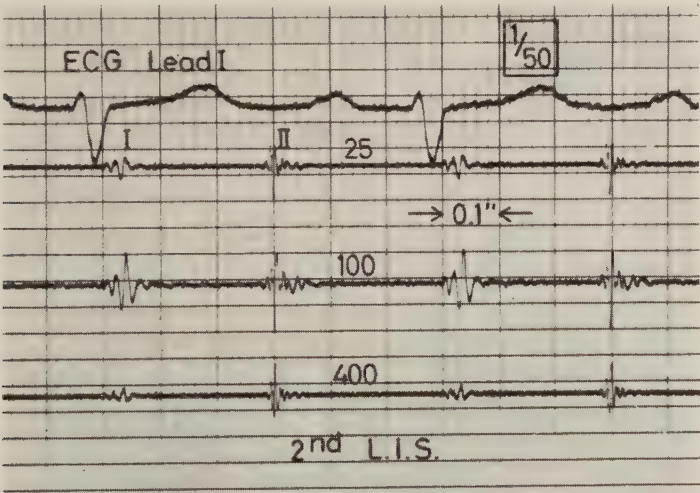


Fig. 8. Phonocardiogram. L. B. 16-year-old boy with tetralogy of Fallot. The heart sounds are clear and no murmur is registered over the second left intercostal space. Autopsy a year later revealed maximal pulmonic stenosis of valvular and infundibular type. The blood flow through the pulmonic ostium presumably had been negligible.

Boxed figures denote degree of amplification. Other figures show standard frequencies of the filters.

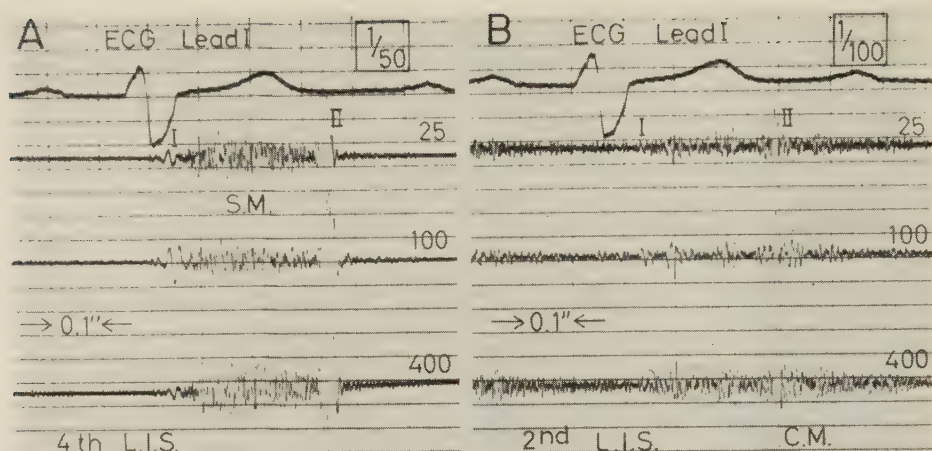


Fig. 9. Phonocardiogram. H. L. 14-year-old boy with tetralogy of Fallot. Infundibular pulmonic stenosis had been demonstrated by angiocardiography.

A. A systolic murmur (S.M.) occurs with maximum amplitude in midsystole. The aortic component of the second sound has large amplitudes. There is no pulmonic component. B. Registration over the second left intercostal space shows a continuous murmur (C.M.) caused by the patent artificial anastomosis.

Boxed figures denote degree of amplification. Other figures show standard frequencies of the filters.

25 patients were dead. As a sign of presumably functioning anastomosis, a continuous murmur was heard, in addition to the systolic murmur, in varying strength over the anastomosis region in 117 cases (86 per cent). (Fig. 9). Phonocardiography showed the systolic murmur to be unchanged by operation in 95.9 per cent of cases (murmur confined to the first half of systole in 26.7 per cent, to midsystole in 52.6 per cent and to late systole in 20.7 per cent). In 4.1 per cent of cases, however, the murmur had shifted from midsystole to early systole.

Of the 8 patients with systolic click, or aortic ejection sound, before operation, 7 still had this sign postoperatively, and in the other case it had disappeared. After operation there were 7 more cases with systolic click, the interval from the first heart sound being 0.06 to 0.08 second. Increase of the minute volume through the aortic valves as a result of the shunt and further dilatation of the proximal part of the ascending aorta were possible causes of this systolic click.

Duplication of the second heart sound as a sign of delayed closure of the pulmonic valve arose postoperatively in 6 cases. The interval between the 2 components of the sound was 0.04 to 0.12 second (mean 0.07 second).

In all these cases there were signs of a well-functioning anastomosis and, having regard to the aetiology, it could be assumed that the pulmonary artery flow was substantially increased by operation (McKusick).

Blood Pressure in the Arms after Blalock Anastomosis

In the anastomosis operations the arterial circulation to the arms was altered only in the cases in which the subelavian artery was used. Cases treated by the Potts direct aorticopulmonary anastomosis or by the Blalock aorticocarotid anastomosis thus showed no significant alteration of the arm blood pressure as compared with the preoperative readings, unless complications had occurred.

For measurement of blood pressure an inflatable cuff and a mercury manometer were used and the measurements were made as recommended by the Swedish Cardilogic Society. The values were the mean of several measurements. Evaluation was based on the systolic readings. A discrepancy of more than 15 mm Hg between both arm pressures was counted as definite difference. The results are shown in table 11.

Fall in the arm blood pressure has no clinical significance with respect to the function in the artificial anastomosis. It is instead an expression of change in the persistent collateral circulation to the arms. In many cases it was found that the blood pressure in the arm of the anastomosis side first became measurable a year or more after operation, which suggested gradual increase in the collateral function.

TABLE 11
Blood Pressure in the Arms after Blalock Anastomosis

	No. of cases
Normal and equal blood pressure in both arms	7
Lower pressure on the side of the anastomosis	51
Non-measurable pressure on the side of the anastomosis	41
Non-measurable pressure in both arms	2 ¹
Pressure record only of contralateral arm	7
No pressure records	15
Dead at time of follow-up	25
Total	148

¹ Subelavian anomaly in 1 case

In no case was operation followed by serious disturbance of circulation which resulted in obvious functional damage to the ipsilateral arm. Both subjectively and objectively some fall in temperature occurred in the arm and hand, and occasionally there was slight impairment of the gross muscular power, particularly in the hand.

CHAPTER 6

ELECTROCARDIOGRAPHY

Introductory Notes

In recent years an abundant literature has demonstrated the great importance of electrocardiography in the diagnosis of congenital heart disease. With the aid of electrocardiograms many problems of differential diagnosis have been solved, not least in the morbus caeruleus group, and prognostic studies have been carried out. SCHMIDT (1961) stated that electrocardiography is more important for haemodynamic analysis than any other method of examination.

Electrocardiographic patterns associated with the tetralogy of Fallot were described by DONZELOT ET AL. (1951), WOODS (1952), SODI-PALLARES & MARSICO (1955), PILEGGI ET AL. (1960), BENDER, DREIFUS & DOWNING (1961) and SCHMIDT.

RIHL, TERPLAN & WEISS (1929) seem to have been the first to publish electrocardiograms in tricuspid atresia with left axis deviation. TAUSSIG (1936) pointed out the diagnostic value of left axis deviation in a cyanotic infant and expressed the opinion that such deviation was a probable sign of tricuspid atresia or hypoplasia of the tricuspid valves. That normal axis may be present in tricuspid atresia, however, was shown by DONZELOT ET AL. (1950), ELSTER (1950), ABRAMS & ALWAY (1951) and KREUTZER ET AL. (1954). In some cases of the same cardiac anomaly, right axis deviation was found (DICKSON & JONES 1948, CHICHE 1952, ASTLEY ET AL. 1953, KREUTZER ET AL., KEITH, ROWE & VLAD 1958).

The electrocardiogram in transposition of the great vessels resembles in many cases that in the tetralogy of Fallot (CAMPBELL & SUZMAN 1951, ASTLEY & PARSONS 1952) but it may also be uncharacteristic, as in cor biloculare and triloculare.

From the electrocardiographic aspect, pulmonic stenosis with normal aortic root shows a varying picture, which was compared with Fallot's tetralogy by SOULIÉ, JOLY, CARLOTTI & SICOT (1951). The importance of right ventricular hypertrophy for the electrocardiogram in pulmonic stenosis was mentioned by CAMPBELL & BROCH (1955) and MARQUIS (1951).

Methods

The electrocardiograms were in most cases registered by means of a six-channel photographically registering apparatus. In a smaller number of cases studied in recent years, a four-channel direct writing apparatus with "jet recorders" (Mingograph) was used. Both types of apparatus were made by Elema (Stockholm) according to Elmqvist's design.

During the period 1947 to 1949 only standard leads were used. After 1949 most cases were studied with standard leads, unipolar limb leads and chest leads according to Wilson from V_1 to V_6 . More recently, V_{4r} and V_7 were also included. As a rule, however, these leads were not available preoperatively. For this reason they are not included in comparisons of electrocardiograms before and after operation. When available, V_{4r} and V_7 were used in the diagnosis of cardiac hypertrophy.

Before operation 56 patients were electrocardiographed with standard leads only. In 102 cases standard leads, unipolar limb leads and chest leads were used preoperatively. In 3 cases operation was not preceded by electrocardiography. Postoperatively 19 patients were studied with standard leads only and 119 with standard, unipolar limb and chest leads. Of these 119 cases, 89 could be compared before and after operation. All recordings were made with the patient in basal state and recumbent position. Much effort was expended to keep the children, particularly the youngest ones, calm and cooperative. No drugs were given in association with electrocardiography, since it was desired to avoid extraneous influence on the record. The time of the latest electrocardiographic follow-up was the same as the latest examination (cf. observation periods in figure 2).

Concerning normal electrocardiographic values, my assessments are based on the values stated by ZIEGLER (1951) for normal infants and children. These norms have long been generally accepted, although many other writers have published normal electrocardiographic standards for children (BURNETT & TAYLOR 1936, HAFKESBRING, DRAWE & ASHMAN 1937, MANNHEIMER 1940, SEHAM & MOSS 1942, EPSTEIN 1948, GROEDEL & MILLER 1944, KOSSMAN 1953, KRUMBHAAR & JENKS 1917, LINCOLN & NICHOLSON 1928, ALIMURUNG & MASSELL 1956, ASHMAN & HULL 1944, LEPESCHKIN 1951, BENGTSSON 1957, SODI-PALLARES ET AL. 1958, HECK & STOERMER 1959, MCCAMMON 1960).

All the electrocardiograms were reviewed and assessed by me.

Cardiac Rhythm

Regular sinus rhythm was present before operation in 152 of the 158

patients examined electrocardiographically. Two patients had sinus arrhythmia with frequency variations of at least 30 beats per minute. Another patient (Case 55) had 2:1 block alternating with extrasystolic arrhythmia (ventricular bigeminally occurring extra systoles) both before and after operation. In Case 36 sinus rhythm alternated with ectopic atrial rhythm patterns preoperatively, but postoperatively the rhythm was normal. Cases 109 and 115 showed ventricular extra systoles before operation. The rhythm normalized postoperatively in the former patient and the latter died at operation. Sinus tachycardia of moderate degree with regard to age was found during basal state in 5 patients preoperatively, and in 2 of these the tachycardia persisted after operation. In the latest post-operative follow-up examinations, sinus arrhythmia was present in 8 cases of 138. In Case 10 the rhythm was normal before operation, but afterwards there were occasional atrial extra systoles without demonstrable cause. In Case 139 a large left-right shunt resulted from excessive width of the artificial anastomosis and, in association with progressive cardiac enlargement and decompensation, there were frequent attacks of atrial paroxysmal tachycardia. Neither before nor after operation was Wolff-Parkinson-White syndrome demonstrable.

P-R Interval

Prolongation of the atrioventricular conduction time seems from the literature to be fairly uncommon in the tetralogy of Fallot. BENDER ET AL. (1961) reported that 5 cases of 100 had first-degree block.

In my series, variations in atrioventricular conduction time in excess of the normal age variations were found in only a few cases. Cases 15, 50 and 77 showed "wandering supraventricular pacemaker" with P-R interval ranging from 0 to 0.18 second. This pattern was present both preoperatively and postoperatively in 2 cases, and in the third there was stabilization of the pacemaker after operation. In 4 patients with normal P-R interval before operation, first-degree block occurred postoperatively, the highest value being 0.30 seconds. In 3 of these patients (Cases 92, 102 and 139), postoperative roentgenograms showed pronounced cardiac enlargement, in all probability attributable to over-dimensioned anastomosis with large left-right shunt. In the fourth case there was no clinically acceptable reason for the first-degree block. According to KEITH, ROWE & VLAD "absence of the segment between the end of the P wave and the beginning of QRS" is a frequent finding in tricuspid atresia. In their series it occurred in 53 per cent of the tracings. Four of my 8 cases of tricuspid atresia showed this picture preoperatively and postoperatively.

P Waves

NADAS (1957) stated that "peaked or gothic-shaped tall, but not prolonged P waves in leads II and III, coupled with pointed P waves in chest leads V_{2-3} , usually indicate right atrial hypertrophy (P pulmonale). In contrast, flat-topped P waves in leads I and II of normal height but prolonged duration, with two notches at least 0.05 second apart, indicate left atrial hypertrophy (P mitrale). In the chest leads prominent and late P waves may be seen in V_5 and V_6 ". According to EAST & BAIN (1959) right atrial hypertrophy is indicated by "diphasic P waves with predominance of the positive phase and with rapid inscription of its intrinsicoid deflection in the right precordial leads". SODI-PALLARES ET AL. (1958) wrote that in general tall P waves suggest enlargement of the right atrium, while broad P waves suggest enlargement of the left atrium, and the presence of both features indicates enlargement of both atria. The tall, early waves are visible in the right chest leads and the late ones in the left chest leads (THOMAS & DEJONG 1954). The diphasic P waves have been called P mitropulmonale by WINTERITZ (1935) and P cardiale by SCHMIDT. This last writer stated that in right atrial hypertrophy the pressure in the right atrium is in proportion to the strength of the atrial wall and the amplitude of the P waves. No clear relationship has been demonstrated between the right atrial pressure and the height of the P waves. Elevation of right ventricular pressure is accompanied by increase in the amplitude of the P waves (MICHEL ET AL. 1957). Reduction of this pressure accordingly leads to diminution of the P waves after a time. This has been observed after successful operations on congenital cardiac malformations with right ventricular strain (LANDTMAN 1954). The importance of arterial anoxaemia for the height of the P waves in right cardiac strain was pointed out by PENALOZA (cited by SODI-PALLARES 1958) and OLIVEIRA & ZIMMERMAN (cited by SCHMIDT 1961).

Right atrial hypertrophy was stated by DONZELOT ET AL. (1951) to occur in one-third of cases of Fallot's tetralogy. In older patients, JOLY, FOLLI & CARLOTTI (1952) found right atrial hypertrophy in one-fifth of cases. KEITH, ROWE & VLAD reported that none of their patients younger than one year showed abnormally tall P waves, and even among the older children abnormal P waves were not common (1:10). Pathologic P waves were found in about 30 per cent of cases of Fallot's tetralogy by MASSIE & WALSH (1960) and in about 50 per cent by BENDER, DREIFUS & DOWNING: In pulmonic atresia and in the "grandes trilogies" the frequency was higher. In tricuspid atresia, right atrial, left atrial or combined forms of

hypertrophy may occur. According to TAUSSIG (1947), the height of the P wave is in indirect relation to the size of the atrial septum defect. This was confirmed by NEIL & BRINK (1955). The frequency of atrial hypertrophy is higher in tricuspid atresia and transposition of the great vessels than in the tetralogy of Fallot or pulmonic stenosis with intact interventricular septum.

In my cases the P waves before operation were not pathologically wide in standard leads or in unipolar precordial or limb leads. After operation broadened P waves were found in only 2 patients (Cases 102 and 139), with the highest value in standard lead II (0.10 and 0.14 seconds). In both cases there was considerable postoperative cardiac enlargement with hypertrophy and dilatation, especially of the atria. The amplitude of the P waves is surveyed in table 12. From the cases studied with lead V₁, those showing diphasic P waves have been deducted in this table.

T A B L E 12
Amplitude of P Waves Before and After Operation

Mean in 1/10 mV	All cases	n	Cases with preop. and postop. readings	n
In lead II before operation	3.1	158	3.1	135
„ „ II after „	2.8	138	2.8	135
„ „ V ₁ before „	2.3	102	2.4	89
„ „ V ₁ after „	1.7	119	1.9	89
„ „ V ₅ before „	1.7	102	1.7	89
„ „ V ₅ after „	1.7	119	1.6	89
„ „ V ₆ before „	1.6	102	1.5	89
„ „ V ₆ after „	1.4	119	1.4	89

Between the total series and the group in which preoperative and postoperative tracings could be compared, there was good conformity as regards mean amplitude of P waves.

Statistical analysis of P-wave amplitude in lead V₁ was made in cases with such readings both before and after operation. From the total 89 cases, 30 with diphasic P waves were excluded. In each of the remaining 59 cases the preoperative value was compared with the postoperative value. A small diminution of P-wave amplitude was then found postoperatively in lead V₁, but it was not statistically significant. The mean P-wave amplitude was calculated for all cases examined with lead V₁ (after exclusion of the cases with diphasic P waves). The values for the 98 preoperatively and the 72 postoperatively examined cases were then distributed according to age of the patients.

	3—5 yrs	5—8 yrs	8—12 yrs	> 12 yrs
<i>Mean in 1/10 mV</i>				
Before operation	2.63	2.81	2.33	1.90
After operation	2.67	2.70	2.30	1.97
<i>No. of cases</i>				
Before operation	41	31	21	5
After operation	8	24	25	15

No significant change in the P-wave amplitude was found in the total series or in the various age groups.

Comparative analysis of the individual changes in amplitude of the P waves in lead V_5 was carried out in the 89 cases with both preoperative and postoperative readings. No significant change was found. The mean preoperative and postoperative values for P-wave amplitude in the same lead were calculated for all the cases so examined (102 before and 103 after operation). These cases were also distributed according to age.

	3—5 yrs	5—8 yrs	8—12 yrs	> 12 yrs
<i>Mean in 1/10 mV</i>				
Before operation	1.77	1.80	1.38	1.20
After operation	1.54	1.77	1.58	1.67
<i>No. of cases</i>				
Before operation	41	35	21	5
After operation	12	28	36	27

No significant change was found, although there was some tendency to increase in the older patients and some decrease in the younger patients.

Individual comparisons were further made of the P-wave amplitude in lead V_6 before and after operation (89 cases). Clearly significant diminution was found. The same tendency appeared when the total 102 examined cases were distributed according to age, though significance was not demonstrated.

	3—5 yrs	5—8 yrs	8—12 yrs	> 12 yrs
<i>Mean in 1/10 mV</i>				
Before operation	1.65	1.54	1.43	1.30
After operation	1.46	1.36	1.24	1.36
<i>No. of cases</i>				
Before operation	41	35	21	5
After operation	12	28	36	26

The reason for the significant decrease of P-wave amplitude in lead V_6 could not be conclusively established. Appreciable reduction of the muscle mass of the left atrium after creation of the anastomosis and the resultant increase of the blood flow from the lungs seems to be unlikely. Some dilatation of the left atrium by the augmented circulation cannot be excluded, and this might produce a fall in P-wave amplitude. Definitive elucidation of the reduced amplitude of the P-waves would seem to be scarcely feasible by electrocardiography.

Table 13 shows the distribution of normal and pathologic P waves among the various diagnosis groups (normal values according to ZIEGLER). Standard lead II was used in all 135 cases examined both before and after operation. In 89 cases Wilson leads V_1 , V_5 and V_6 were used for study of the atria. It is seen that the greatest postoperative variation occurred in the

TABLE 13
*Frequency of Normal and Pathologic PWaves Before and After Operation Distributed
 According to Diagnosis*
 (135 cases with standard leads both before and after operation
 89 cases with chest leads both before and after operation)

Diagnosis		Lead II		V_1		V_5		V_6	
		preop.	postop.	preop.	postop.	preop.	postop.	preop.	postop.
Tetralogy of Fallot	n	85	82	40	39	33	37	39	54
	p	39	42	41	42	48	44	42	27
Tricuspid atresia	n	0	3	3	2	1	0	1	0
	p	6	3	1	2	3	4	3	4
Transposition	n	1	1	1	1	0	1	0	0
	p	1	1	0	0	1	0	1	1
Pulmonic stenosis with ostium atr. ventr. com.	n	0	1	1	1	0	1	0	1
	p	1	0	0	0	1	0	1	0
Pulmonic stenosis with atrial septal defect	n	1	1	0	0	0	1	1	1
	p	0	0	1	1	1	0	0	0
Diagnosis not clear	n	0	1	1	1	0	0	0	0
	p	1	0	0	0	1	1	1	1
Total number	n	87	89	46	44	34	40	41	56
	p	48	46	43	45	55	49	48	33
per cent	n	64.4	65.9	51.7	49.4	38.2	44.9	46.1	62.9
	p	35.6	34.1	48.3	50.6	61.8	55.1	53.9	37.1

n = normal
 p = pathologic

left precordial leads, in which there was appreciable normalization of the P waves. No significant change occurred over the right side of the precordium.

QRS Interval and Axis

The QRS interval in standard lead II was determined in 158 cases before shunt operation and in 137 cases postoperatively. Normal values (respective means 0.061 and 0.066 second) were found in all but 2 patients (Cases 17 and 139) with tetralogy of Fallot. In Case 17 the QRS interval was 0.10 second before operation and 0.12 second afterwards; no clear explanation was found for these relatively high values. In Case 139 the QRS interval increased from 0.04 before to 0.14 after operation. The probable cause was excessive width of the anastomosis which resulted in a large left-right shunt with progressive cardiac dilatation and hypertrophy.

The QRS axis is one of the standard items of clinical electrocardiography. WALLER (1913) was the first to use the term "electrical axis". ASHMAN & HULL (1944) stated that the mean or average electrical axis of the QRS complex is "ordinarily determined" by the algebraic sum of the amplitudes of R and S in leads I and III, but that for the correct determination the net area (algebraic sum of the areas of upward and downward deflection) should be used. The difference when the axis is determined from the net area and from the net amplitude is relatively small, and determination of axis from algebraic sums of deflection is now generally accepted. This is the method I have used for determining electrical axis in the present study.

The electrical axis before operation varied between -5 and $+65$ degrees and after operation between -5 and $+80$ degrees (fig. 10). The mean value for the electrical axis preoperatively (158 cases) was $+135.7$ and at the latest postoperative follow-up it was $+132.8$ degrees (137 cases). The 134 cases in which the electrical axis could be compared preoperatively and postoperatively are presented according to diagnosis in table 14. The distribution seemed to be in good conformity with the values stated by SODI-PALLARES ET AL. (1958) for "mean manifest electrical axis of the ventricular activation process in congenital heart disease".

In statistical analysis with respect to changes in position of the electrical axis, individual comparisons of preoperative and postoperative readings were made in 121 selected cases. In order to obtain a statistically homogeneous material, no case with extreme axis position was included. A clearly significant difference was found after operation. The mean value for all 121 cases was $+121.8$ degrees before operation and $+116.7$ degrees after

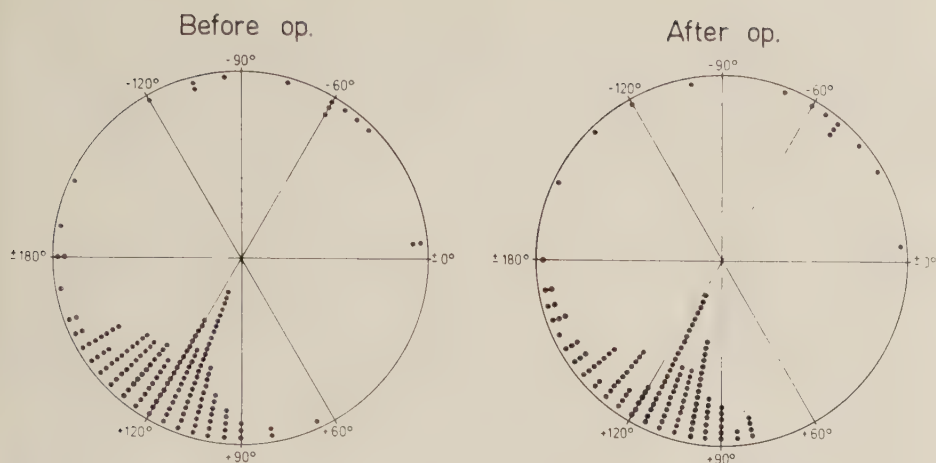


Fig. 10. The position of the electrical axis in 158 cases before and 137 cases after shunt operation. Each dot represents a case.

TABLE 14
The Electrical Axis Before and After Operation
Distributed According to Diagnosis
(134 cases with values both before and after operation)

Diagnosis	No. of cases	Mean electrical axis of QRS		difference
		preoperative	postoperative	
Tetralogy of Fallot	123	+129.7°	+125.6°	— 4.1°
Tricuspid atresia	6	— 76.7°	— 71.0°	— 5.7°
Transposition	2	+170°	+177°	+ 7°
Pulmonic stenosis with ostium atrioventriculare commune ...	1	+170°	+140°	—30°
Pulmonic stenosis with patent foramen ovale	1	+ 90°	+105°	+15°
Diagnosis not clear	1	+105°	+100°	— 5°
Total	134	+137.0°	+133.4°	— 3.6°

operation. A 95 per cent confidence interval gave a range of +2.4 to -7.7, with a mean of 5 degrees.

When the cases were distributed according to age, the following means were obtained.

	3—5 yrs	5—8 yrs	8—11 yrs	12—16 yrs
Mean values (degrees)				
Before operation	123.6	122.8	123.1	116.3
After operation	121.0	122.8	117.3	112.2

The decrease in right axis deviation after operation was most pronounced in the older patients. Thus, in the patients aged 8 to 12 years there was a reduction of 5.8 degrees and in those aged 12 to 16 years the decrease was 4.1 degrees. The corresponding values in the 3 to 5 and 5 to 8 age groups were 2.6 degrees and nil.

R Waves

If the R wave in lead V_1 is regarded as an expression of the action potential of the right ventricle, a postoperative change in this R wave should denote a change in the activity of the right ventricle. Comparisons between preoperative and postoperative values must be made on cases with a similar anatomic pattern, in which the function of the ventricles is of the same type (cf. the tetralogy of Fallot with right ventricular hypertrophy and tricuspid atresia with non-functioning right ventricle). Of the 89 cases in this series which were studied with unipolar precordial leads both before and after operation, 84 had tetralogy of Fallot. The remaining 5 cases with other cardiac anomalies constituted too small a group for statistical analysis.

The amplitude of the R waves in lead V_1 in the 84 cases of Fallot's tetralogy showed no statistically significant change when individual comparisons were made between preoperative and postoperative readings.

If the R wave in leads V_5 and V_6 is regarded as an expression of the action potential of the left ventricle, postoperative change in these R waves should indicate altered left ventricular activity. In the 84 cases of the tetralogy of Fallot which were compared in the same way as for lead V_1 , a statistically significant increase was found postoperatively in both V_5 and V_6 .

The R-wave amplitude in all the cases of tetralogy of Fallot, distributed as means for different age groups, was compared with the normal values stated by ZIEGLER.

<i>Lead V_1</i>	<i>3—5 yrs</i>	<i>5—8 yrs</i>	<i>8—12 yrs</i>	<i>12—16 yrs</i>
<i>Mean in 1/10 mV</i>				
Before operation	19.4	22.9	18.3	20.5
After operation	20.5	24.6	23.0	22.6
According to ZIEGLER	7.8	7.6	6.5	5.6

Although in all the age groups there was a clear tendency to increase of R-wave amplitude in lead V_1 , the differences were not statistically significant. As compared with the normal values reported by ZIEGLER, all the readings in my patients were pathologically high in this lead, illustrating the right ventricular hypertrophy which is present in the tetralogy of Fallot.

<i>Lead V₅</i>	<i>3-5 yrs</i>	<i>5-8 yrs</i>	<i>8-12 yrs</i>	<i>12-16 yrs</i>
<i>Mean in 1/10 mV</i>				
Before operation	10.2	10.0	11.4	7.8
After operation	17.6	15.4	19.3	20.8
According to ZIEGLER	19.8	21.8	22.8	18.1

In lead V₅ the results of analysis according to age were similar to those in lead V₁, i.e., a distinct tendency to postoperative increase in the amplitude of the R waves, but no clear statistical significance. The fairly large variations in the postoperative values were possibly the reason why the test failed to reveal significant differences between means. In the age group 8 to 12 years the postoperative mean was significantly higher than the preoperative on the 95 per cent level, but not on the 99 per cent level. Increased R-wave amplitude in these age groups thus was more probable in lead V₅ than in lead V₁. ZIEGLER's normal values for R waves in lead V₅ were higher than the values in my cases, except in the patients aged 12 to 16 years.

<i>Lead V₆</i>	<i>3-5 yrs</i>	<i>5-8 yrs</i>	<i>8-12 yrs</i>	<i>12-16 yrs</i>
<i>Mean in 1/10 mV</i>				
Before operation	6.1	5.8	7.9	7.8
After operation	11.8	10.0	10.0	11.8
According to ZIEGLER	13.0	14.3	13.8	13.8

When the values for R-wave amplitude in lead V₆ were grouped according to means in the various age groups there was, as in lead V₅, a tendency to postoperative increase which was not statistically significant. The R waves in lead V₆ in ZIEGLER's normal subjects were distinctly higher than those in my cases. The relatively low amplitude of the R waves in leads V₅ and V₆ may be regarded as indicative of lower left ventricular activity in the tetralogy of Fallot than in healthy individuals. After shunt operation the activity in the left ventricle increased but still was less than normal.

Another feature of the R waves was studied. This was the intrinsicoid deflection, which is also called the "ventricular activation time". It is calculated as the time interval between the beginning of the Q wave and the peak of the latest R wave (R or R') in any one lead. With unipolar precordial leads over the ventricles, it has been theoretically calculated that the ventricular activation time provides some measure of the thickness of the ventricular wall. The normal activation time for the right ventricle has been estimated as less than 0.03 second and the normal time for the left ventricle as less than 0.04 second.

Analysis of the ventricular activation time in my cases showed no significant postoperative increase in lead V_1 (0.035 before and 0.038 after operation). This would seem to indicate that the right ventricular hypertrophy was unaltered by shunt operation in these patients with tetralogy of Fallot. The ventricular activation time over the left ventricle (lead V_6) was significantly increased after operation by 0.006 ± 0.001 second. The mean preoperative value was 0.022 second and the mean postoperative value was 0.027 second. The mean increase in ventricular activation time in lead V_6 was found to be about 25.1 per cent. The results of this investigation thus were in conformity with those in the analysis of amplitude, viz., that the activity of the left ventricle is increased after operation, while that of the right ventricle is unaltered.

Ventricular Hypertrophy

In assessing ventricular hypertrophy in the electrocardiograms, I applied the principles stated by DONZELOT, MÉTIANU & DURAND (1952), KEITH, ROWE & VLAD (1958), GOODWIN (1960) and SCHMIDT (1961) for judging right ventricular hypertrophy, and by SCOTT, SEIWERT, SIMON & MCGUIRE (1955), SELZER ET AL. (1958), KEITH, ROWE & VLAD; CHOU ET AL. (1960) and SCHMIDT (1961) for judging left ventricular hypertrophy.

Of the 102 cases which were examined preoperatively with standard leads and unipolar limb and chest leads, 95 showed signs of right ventricular hypertrophy. Three patients (2 with tricuspid atresia and 1 with reduced pulmonary circulation of obscure causation) had signs of left ventricular hypertrophy.

After operation 119 patients were examined in the same way. In 113 the electrocardiograms suggested right ventricular hypertrophy and in 4 left ventricular hypertrophy (3 with tricuspid atresia and 1 with reduced pulmonary circulation of obscure causation). In 2 cases there were no signs of hypertrophy (1 with tricuspid atresia and 1 with tetralogy of Fallot).

Of the 8 cases in which the clinical diagnosis was tricuspid atresia or stenosis, 2 could not be judged with respect to ventricular hypertrophy before or after operation, as they were not examined with unipolar chest leads. Two patients had left ventricular hypertrophy preoperatively and postoperatively. Three patients had no ventricular hypertrophy before operation: 1 of them had postoperative left ventricular hypertrophy and 2 died at operation. The eighth patient had right ventricular hypertrophy before and after operation, although the haemodynamic findings most

closely corresponded to tricuspid atresia. Autopsy, performed after revision of the shunt operation, showed that this patient had both tricuspid stenosis and pulmonic stenosis and also a patent foramen ovale and intact inter-ventricular septum.

As already stated, electrocardiographic signs of right ventricular hypertrophy were absent in a case of Fallot's tetralogy both before and after operation, although the clinical picture otherwise supported the correctness of the diagnosis. According to TAUSSIG (1960), one need not doubt the diagnosis in such cases. MASSIE & WALSH (1960) found normal electrocardiograms and vectorcardiograms in about 3 per cent of cases of tetralogy of Fallot.

Combined left and right ventricular hypertrophy was not found on the electrocardiograms in any of my cases before or after operation. In 2 cases of tetralogy of Fallot the electrocardiograms were characteristic of mirror image or true dextrocardia, with right ventricular hypertrophy which was unaltered after operation. No patient with tetralogy of Fallot had left ventricular hypertrophy, which has been stated to be rare in the tetralogy (GASUL ET AL. 1949, ZIEGLER 1954, KEITH, ROWE & VLAD).

RS-T Segment

This segment is the interval between the end of the QRS complex and the beginning of the T wave. Depressions of the segment by 1.5 mm or more were rare in this series of patients. In 4 patients with tetralogy of Fallot, who had severe right ventricular hypertrophy, appreciable depression of the RS-T segment was observed in the right precordial leads. This was interpreted as a sign of right ventricular strain. No distinct abnormality of the segment was observed in the left precordial leads before or after operation.

T Waves

The T wave, the terminal portion of ventricular activity, shows wide variations among patients with the same cardiac anomalies. According to TAUSSIG (1960), the T waves may or may not be abnormal. Because of the possible variations, it does not seem feasible to state firm rules for the occurrence of T-wave changes in congenital cardiac disease. In my series, 89 cases (for diagnoses see table 13) were compared before and after operation with respect to T waves. The number of inverted T waves was significantly increased after operation in all unipolar precordial leads.

This increase was most pronounced in leads V_1 and V_2 . The change may have been attributable to the anastomosis operation and the resultant cardiovascular changes, or it may have been referable to the ordinary development of the electrocardiogram in regard to age and pathologic anatomy.

Q—T Interval

The Q—T interval measures the total electrical systole of the ventricles. When the corrected Q—T interval was calculated from actual measurements and cycle length in seconds using BAZETT's (1920) formula, the graph published by ALIMURUNG, JOSEPH, CRAIG & MASELL (1950) and calculations by ZIEGLER (1951), no definitely pathologic values were found. Shunt operations thus did not seem to have significant effect on the Q—T interval.

Summary

1. Of 161 patients with morbus caeruleus, 158 were examined by electrocardiography before operation. In 56 cases only standard leads were used, and 102 cases were studied with standard leads and unipolar limb and chest leads. Postoperative electrocardiography was done in 138 cases—with standard leads in 19 and with standard leads and unipolar limb and chest leads in 119 cases. Of these 119 cases, 89 had been examined in the same way before operation.
2. Arrhythmia with extra systoles was uncommon after shunt operation, only 1 patient being found to have atrial extra systoles. In another case frequent attacks of paroxysmal tachycardia developed in association with progressive cardiac enlargement and decompensation, probably as a result of excessive width of the artificial anastomosis.
3. Prolonged atrioventricular conduction time (P-R interval) was found postoperatively in 4 cases, 3 of which showed roentgenologic cardiac enlargement after the shunt operation.
4. Broadening of the P waves occurred in only 2 patients postoperatively. By contrast, the amplitude of the P waves altered in that significant decrease was found postoperatively over the left side of the heart. No significant change in the P waves was found over the right side of the heart.
5. The mean value for the electrical axis of the QRS complex showed significant diminution in right deviation after operation.

6. The R waves over the left side of the heart showed significant increase of amplitude after operation in the patients with tetralogy of Fallot, whereas the R waves over the right side of the heart were not significantly altered. The increased R-wave amplitude in the left precordial leads is attributed to the increase in the work of the left ventricle following creation of the anastomosis.
7. Hypertrophy of the right ventricle was present in almost all of the series. Exceptions were patients with tricuspid atresia and a case in which the diagnosis was obscure: These patients had no or only left ventricular hypertrophy. One patient with tetralogy of Fallot had no definite hypertrophy before or after operation, which is considered to be unusual in view of the diagnosis.
8. Significant increase in the number of inverted T waves was noted after operation in the unipolar precordial leads. This change was most pronounced over the right side of the heart. Its cause could not be conclusively established.

CHAPTER 7

HEART CATHETERIZATION

Introductory Notes

Ever since FORSSMAN (1929) first attempted venous catheterization of the heart, this form of investigation has undergone constant improvements. The work of Cournand & Ranges (1941) attracted general interest to heart catheterization. The elaborations of Cournand (1949), Bing (1947) and Dexter (1947) gave the technique essentially its present character. In the past decade heart catheterization, because it permits measurements of pressure and blood oxygen saturation within the heart, has proved exceedingly helpful in the diagnosis of congenital cardiac malformations. Children of all ages have been so examined in steadily increasing numbers, as witnessed by the voluminous literature.

In my series of patients with morbus caeruleus, the anatomic circumstances frequently were highly complicated by the multiplicity of malformation. This impeded heart catheterization in many cases. In the patients with the tetralogy of Fallot, for instance, it was very difficult to reach the pulmonary artery *via* the right ventricle if the pulmonary stenosis was pronounced. Consequently, the pulmonary artery pressure before and after shunt operation could be registered only in a small number of patients. In cases of tricuspid atresia, satisfactory catheterization likewise was difficult. In no case of this type could the pulmonary artery be reached with the catheter. The poor general condition of most of the children with morbus caeruleus was also to some extent a hindrance to these exacting investigations. When the pulmonary stenosis was very severe, passage of the catheter into the pulmonary artery could produce attacks of cyanosis and dyspnoea so grave as to endanger life. The pressure in the pulmonary artery thus was influenced by the degree to which the catheter obstructed the blood flow from the right ventricle to the pulmonary artery in the stenosed region. The position of the catheter in relation to the flow within the stenosed pulmonary artery was another factor of importance for the pressure.

Methods

Heart catheterization was performed according to the technique described by Cournand, Baldwin & Himmelstein (1949), Lagerlöf, Mannheimer & Werkö (1949) and Ziegler (1954). Cournand catheters and, later on, also Lehman catheters were used in sizes from 5 to 9. The pressure registrations between 1947 and 1953 were made with a Tybjaerg Hansen (1949) condenser manometer (Kaiser Laboratories, Copenhagen). After 1953 a strain gauge manometer in combination with a carrier-frequency amplifier (Elema, Stockholm) was used. Holmgren (1956) has studied the frequency response of the manometer. With both instruments the conditions for investigation were the same. As zero level was selected the atmospheric pressure on a plane 2 cm dorsal to *angulus Ludovici* in infants and very young children and 5 cm dorsal to this angle in older children and adults. Before and after each pressure recording the zero level and a known calibrated standard pressure were registered. The standards were calibrated against a water or mercury manometer. A six-channel photographic electrocardiograph (Elema, Stockholm) registered the tracings. In some early cases at Sabbatsberg Hospital, pressure was measured with a water manometer and only mean pressure was registered. Normal values (mm Hg) for pressures in right cardiac catheterization have been stated by Kjellberg, Mannheimer, Rudhe & Jonsson (1959).

Blood for analysis of oxygen content was sampled through the catheter. The method of van Slyke (1924) was used up to the end of 1953. Thereafter most of the analyses were made with a Brinkman haemoreflexor (Kipp, Delft). This method was described by Zijlstra (1951). The tests with the Brinkman apparatus were checked by analyzing random samples according to van Slyke. Since the van Slyke tests were more accurate but required twice as much blood as the Brinkman method, double analyses were seldom made in the children so tested. For Brinkman analyses, on the other hand, the smallness of the necessary blood quantities permitted at least double specimens to be taken from each sampling site. The systemic arterial oxygen saturation was measured from samples taken during heart catheterization, as a rule from the femoral artery, but in some of the older children and adults from the brachial artery. In many cases in which the catheter could be guided into the over-riding aorta, blood from the descending aorta was taken for measurement of oxygen saturation. All these samples were analyzed with the van Slyke method.

Estimation of the minute volume of blood according to Fick's principle was not carried out, the reason being the many sources of error in right-

left shunt. The blood flow through the pulmonary artery is small, but the existence of collateral circulation *via* the bronchial arteries precludes use of the Fick principle to calculate pulmonary artery flow. This was earlier pointed out by BING, VANDAM & GRAY (1947).

As premedication a barbiturate was given, the aim being sedation without influence on circulation or respiration. Only in a few very young or highly apprehensive children was general anaesthesia necessary for catheterization. At the examination the patients were fasting and in as basal a state as their age permitted.

Results

In 101 cases preoperative heart catheterization was carried out. Postoperatively 89 patients underwent heart catheterization after varying observation periods. Since great importance was attached to pressure and flow in the pulmonary artery, it may be mentioned that the catheter was introduced into this vessel in 42 cases prior to operation and in 44 cases after operation. Only in 16 cases of Fallot's tetralogy was the main trunk of the pulmonary artery entered by the catheter both before and after creation of shunt. The pressure measurements in these cases are presented in table 15.

The preoperative systolic pressure in the pulmonary artery did not exceed 15 mm Hg in 11 cases (table 15) and thus may be characterized as subnormal due to the pulmonic stenosis. The highest pressure in the group was 25 mm Hg in one patient. Following the operation only 4 patients had systolic pulmonary artery pressure lower than 15 mm Hg.

Cases 3 and 146 had pulmonary arterial hypertension postoperatively; the respective pressures in the main pulmonary artery trunk were 40/10 (mean 27) mm Hg and 37/10 (mean 22) mm Hg. In both cases there were clinical signs of a functioning shunt, including a continuous murmur that had appeared postoperatively over the site of the anastomosis. The physical powers of both patients were remarkably good after the operation. The oxygen saturation of the peripheral arterial blood was high in consideration of the cardiac anomaly—c. 90 to 91 per cent during rest in both cases. That the cause of the hypertension was a large left-right shunt through the Blalock anastomosis, with resultant increase in pulmonary flow, seemed likely with regard to the findings at follow-up examination. In both cases the PCV pressure was difficult to assess for calculation of pulmonary resistance. The pulmonary hypertension has not yet become severe enough to produce symptoms from the patent anastomosis. The heart volume was normal in both cases.

TABLE 15

Cases with Preoperative and Postoperative Pressure Measurements

Case no. Sex	Age at operation (years)	Pressure in mm Hg						Postopera- tive observa- tion (years)
		Before operation			After operation			
		R. ven- tricle	Pulmon. artery	Aorta	R. ven- tricle	Pulmon. artery	Aorta	
3 ♀	3	84/3	25/7	—	100/12	40/10	—	4
9 ♂	5	88/3	10/3	—	100/0	13/8	98/57	7
10 ♀	6	70/17	10/6	100/66	112/0	19/13	117/75	7
38 ♀	8	112/0	17/7	112/74	112/0	20/5	105/70	5
69 ♀	13	140/0	18/10	—	114/10	23/14	—	5
87 ♀	12	88/0	14/7	83/54	100/0	16/11	100/64	6
93 ♂	12	100/0	7/3	—	122/0	19/6	—	4
97 ♀	5	95/6	7/3	95/72	109/3	14/9	—	4
113 ♀	4	80/7	19/12	85/54	107/5	16/11	—	3
117 ♀	10	70/10	7/4	95/80	98/0	18/9	98/70	7
126 ♀	3	80/0	12/9	—	100/0	22/9	—	9
131 ♂	7	>50	12/7	—	95/0	17/7	97/58	7
145 ♀	11	91/0	8/6	—	92/0	14/10	—	2
146 ♂	12	85/7	15/5	113/79	110/0	37/10	—	2
157 ♂	1	83/7	22/5	78/47	109/0	20/7	—	1
158 ♀	3	88/5	M 7	—	109/0	14/11	105/73	2

Of the 16 patients with both preoperative and postoperative registration of pulmonary artery pressure, only 1 (Case 126) displayed clinical and roentgenographic signs of nonfunction of the anastomosis due to thrombosis. No continuous murmur could be detected by auscultation or phonocardiography. At follow-up examination there was severe cyanosis and polyglobulia (haemoglobin 21.4 g per cent, erythrocytes 7.5 m. per cu. mm). The oxygen saturation in the femoral artery was 51 to 56 per cent. The pulmonary artery pressure in this patient was both preoperatively and (almost 10 years later) postoperatively within the normal range of variation. The somewhat higher postoperative reading may have been produced by the development of collateral blood channels with the passage of years.

Fifteen of the 16 cases in which pulmonary artery pressure was registered both preoperatively and postoperatively were suitable for statistical analysis. In the other case only the mean pressure was recorded preoperatively, which precluded comparison with the postoperative reading. Because of the smallness of the group, the conclusions are somewhat uncertain. The systolic pressure was significantly increased postoperatively as compared

with the preoperative values. Calculation of the confidence interval showed that with 95 per cent probability the mean increase was between 3.4 and 10.6 mm Hg. The relative wideness of the limits is attributable to the smallness of the group. The diastolic pressure likewise was significantly increased, the confidence interval showing with 95 per cent probability the increase to be between 1.5 and 4.5 mm Hg. Statistical testing of the difference between systolic and diastolic pressure did not show significant difference, but there was some tendency to postoperative increase.

Figure 11 presents all the cases in which the pulmonary artery pressure was registered before shunt operation. They are arranged in order of age. There was no association between systolic pressure and age. In no case was the systolic reading higher than 25 mm Hg. In 30 of the 42 cases the pressure did not exceed 15 mm Hg and therefore must be classed as subnormal. The character of the pressure record made the calculations difficult in many cases.

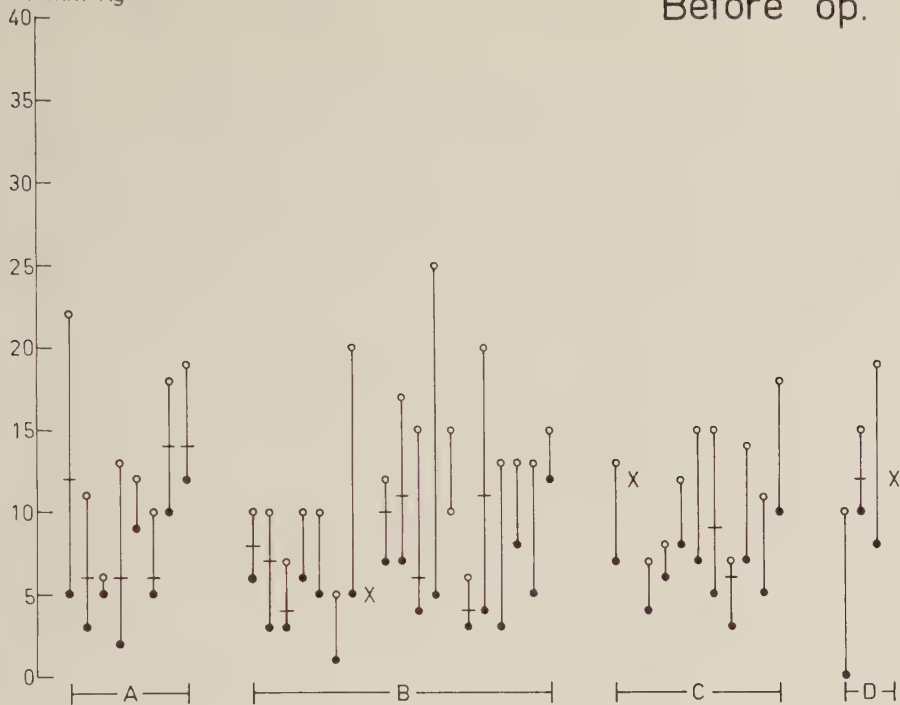
The 44 cases with postoperative measurements of pulmonary artery pressure are shown in figure 11. In 4 cases the systolic reading was more than 30 mm Hg, the highest value being 40 mm. Two of these hypertensive cases have already been discussed, since they also had preoperative measurements. The other 2 cases lacked preoperative values. In all 4 hypertensive patients there were signs of a functioning anastomosis, indicating that the hypertension had developed after the operation as the result of increased blood flow to the lungs *via* the shunt. Roentgenographic evidence of patency was also obtained.

Mean pulmonary artery pressure was measured preoperatively in 19 cases; the average for the group was 9 mm Hg. Corresponding figures for the postoperative investigations were 41 cases and 14 mm Hg. There was thus a rise of 5 mm Hg as compared with the preoperative average.

Since there were only 15 cases with directly comparable preoperative and postoperative measurements of pulmonary artery pressure, it was considered advisable to supplement the analysis by study of all the available pulmonary artery pressure readings before and after operation. The values thus were not all paired readings in the same persons, and individual deviations could be decisive. Although statistically significant difference was not demonstrated, the test showed it to be highly probable that the systolic and diastolic pressures rose after operation.

When the catheter entered the pulmonary artery, an attempt was always made to guide it through the anastomosis. Because of technical difficulties, however, the catheter reached the aorta only in a few cases. The pressure readings and blood gas analyses in all these cases bore witness to the existence of a left-right shunt.

Pressure
in mm Hg



Pressure
in mm Hg

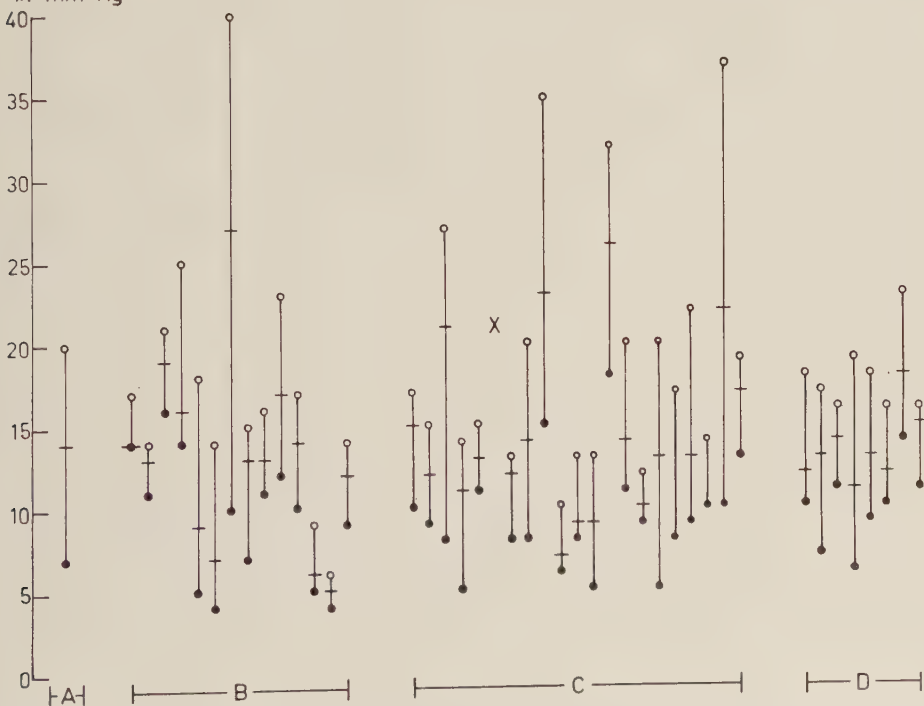


Fig. 11. Pressure in the pulmonary artery before operation (42 cases) and after operation (44 cases). The unfilled circles represent systolic pressure and the filled circles diastolic pressure. The horizontal strokes on the connecting lines (pulse pressure) denote mean pressure.

X = mean pressure measured with water manometer. The cases are divided according to age.

A = ≤ 5 years B = 5–10 years C = 10–15 years D = 15–20 years

Because of the aortic over-riding, the pressure in the right ventricle and that in the aorta were by and large mutually similar during systole in patients with Fallot's tetralogy. The ventricular and aortic pressure measurements in these cases were made separately, not simultaneously. Variations in right ventricular pressure occurred in the Fallot group, but were on the whole insignificant both preoperatively and postoperatively. The pressure in the right atrium was within normal limits except in 11 cases of tetralogy of Fallot and 1 case of tricuspid atresia, in which the a -wave varied between 10 and 17 mm Hg. Mean pressure was not registered. In 3 cases of Fallot's tetralogy the atrial pressures normalized postoperatively. Because of the considerable variations that often are normally found in measurements of interatrial pressure, these values permitted no definite conclusions. It should be noted, however, that in a patient with tetralogy of Fallot (Case 139) the pressure in the right atrium was as high as 17 mm Hg. This was easily explained by the extreme cardiac enlargement and tricuspid insufficiency that had resulted from an excessively wide anastomosis with a powerful left-right shunt. Apart from this case, the shunt operation did not seem to have any noteworthy effect on the pressures in the right cardiac chambers. In 3 of the 8 cases with tricuspid atresia that were catheterized postoperatively, there was slight hypertension in the superior vena cava and the right atrium.

As mentioned, determinations of systemic arterial oxygen saturation were made on blood sampled during heart catheterization. In 69 cases there were comparable preoperative and postoperative values. Figure 12 shows that after operation there was a clear shift of the preoperatively low saturation values towards higher readings. In the same diagram the haemoglobin values are shown in relation to the various levels of systemic oxygen saturation. The mean preoperative and postoperative haemoglobin values in the respective groups were similar, except in one case. Here a single determination showed strikingly low haemoglobin when the arterial oxygen saturation was less than 50 per cent, but this could be ascribed to coexistent disease (pulmonary tuberculosis). Otherwise there was a clear tendency to reduction and normalization of haemoglobin readings as arterial oxygen saturation improved. During the first period of the investigation, from 1947, a standardized Autenrieth haemoglobinometer was used, which gave somewhat uneven readings in single determinations. In order to reduce this source of error, a mean of several determinations was used as the haemoglobin reading in individual cases with multiple tests. Later in the investigation period, haemoglobin was determined by means of a simple filter colorimeter according to Ljungberg.

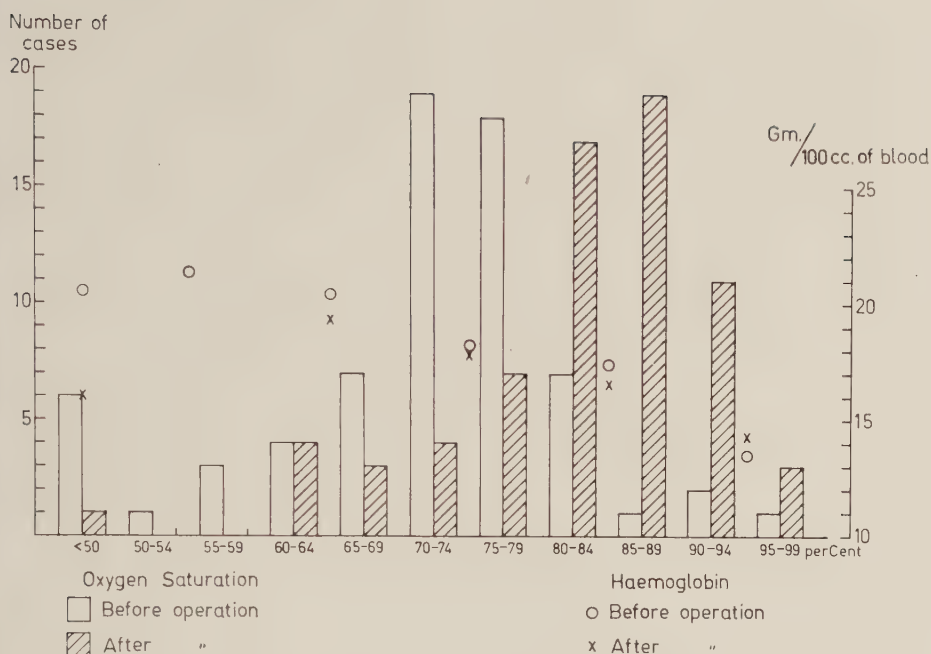


Fig. 12. Arterial saturation in the descending aorta or the femoral artery correlated to haemoglobin before and after operation in 69 comparable cases. The haemoglobin is stated as the mean of all determinations in the relevant O_2 saturation group.

I also compared the erythrocyte count per cu. mm with the arterial oxygen saturation and found the same relation as in the comparisons with haemoglobin. When the oxygen saturation of the systemic arterial blood was low, the erythrocyte count was high, and when the saturation rose the erythrocyte count diminished. The relation between haemoglobin and erythrocyte count seemed to be linear in these cases. In conclusion, it may be stated that polyglobulia showed a clear tendency to diminish as the arterial oxygen saturation rose.

Summary

1. Heart catheterization was performed before shunt operation for morbus caeruleus in 101 cases. Only in 42 of these was it possible to guide the catheter into the pulmonary artery in order to measure the pressure there. After operation 89 cases were studied by heart catheterization and in 44 cases the pulmonary artery pressure could be measured. The difficulties in reaching the pulmonary artery arose from the anomalous anatomy.

2. In 15 cases pulmonary artery pressure could be compared before and after operation. Preoperatively the readings were normal or subnormal. Postoperatively there was statistically significant increase in the systolic and diastolic pressures. These were still within the limits of normal, however, except in 2 patients with slight to moderate hypertension, probably as the result of the increased flow to the pulmonary artery *via* the anastomosis.
3. Among the postoperatively catheterized patients without comparable preoperative measurements, 2 had slight pulmonary hypertension; the systolic pressure did not exceed 40 mm Hg. Comparison between *all* the preoperative and all the postoperative readings of pulmonary artery pressure showed probable increase of the mean systolic and diastolic pressures, but the difference was not statistically significant. The normal limits of variation were not exceeded. Only one patient with hypertension and clinical signs of functioning anastomosis revealed appreciable cardiac enlargement in the roentgenograms.
4. The systemic arterial oxygen saturation could be compared preoperatively and postoperatively in 69 cases. The saturation tended to be distinctly higher after operation, and in some cases the readings during rest were within the limits of normal variation.
5. Polyglobulia was found to be related to systemic arterial oxygen saturation and diminished as the oxygen saturation increased.

CHAPTER 8

ROENTGENOGRAPHY AND ANGIOCARDIOGRAPHY

The roentgenologic studies of the heart in my series of patients included as a rule both fluoroscopy and radiography. Fluoroscopy was valuable for study of certain roentgenologic details and for diagnosis (TAUSSIG 1947). By comparisons of preoperative and postoperative roentgenograms, changes in the size and configuration of the heart and in the vascularization of the lungs could be observed. The examinations were made with the patients in sitting or standing position. Only very young or severely ill children were examined in supine position. Frontal, lateral and oblique views were taken. The volume of the heart was calculated with the aid of the Rohrer-Kahlstorf formula according to the methods used in Sweden. These were described by LILJESTRAND, LYSHOLM, NYLIN & ZACHRISSON (1939) and were modified by JONSELL (1939). Measurements of the cardiac volume in infants were reported by AXÉN & LIND (1945) and LIND (1951). Calculation of body surface area was made according to the Du Bois formula. Normal values of cardiac volume per square metre of body surface area in children were stated by CARLGREN & EEK (1949), KJELLBERG, LIND, LÖNROTH & RUDHE (1954) and LIND. All the roentgenograms of the children in my series, preoperative and postoperative, were uniformly assessed by me and a roentgenologist.

Results of Roentgenography

In all the 161 cases of morbus caeruleus the preoperative roentgenograms showed a common feature, viz., sparsely occurring, narrow and thin pulmonary blood vessels, indicating various degrees of diminution of the pulmonary circulation. Preoperative and postoperative roentgenograms could be compared in this respect in 118 cases. The condition of the pulmonary vascular bed was decidedly improved in 61 cases, the vessels being wider and their image more dense. In 26 further cases there was some,

though less pronounced, postoperative improvement. Among these 87 patients (74 per cent of the 118 cases), the improvement in pulmonary circulation was distinctly greater on the side of the anastomosis in 21 cases. In the remaining 31 of the 118 cases there was no definite improvement in the pulmonary vascular bed. That this roentgenologic assessment of the results of surgery was not in complete conformity with the clinical results was evident from the fact that, of the 31 patients with unaltered roentgenologic picture, only 3 were clinically unimproved. In 4 cases in which the operation result was clinically poor, postoperative roentgenograms showed development of collateral channels. The quality of the films and the roentgenologic technique were less good during the first years of the study than they later became. For this reason, comparisons between preoperative and postoperative films were difficult in many cases and permitted only qualified conclusions.

The roentgenologic picture in most cases of tetralogy of Fallot was characteristic, with selective enlargement and forward bulging of the right ventricle, raising of the apex from the diaphragm and narrowing of the waist of the heart. The pulmonic arch was clearly smaller than normal or was not visualized. Vascular markings in the hilar regions and the periphery of the lungs were scanty. The right atrium was enlarged and the aorta wide in many cases. Although this "cœur en sabot" was the most common picture, variations occurred. Of the 161 cases in the total series, 124 (77 per cent) showed typical Fallot configuration. Of the other cases, 4 (2.5 per cent) showed almost normal roentgenologic image of the heart without distinct enlargement; in 3 of these patients the clinical diagnosis was tetralogy of Fallot and in the fourth it was pulmonic stenosis with atrial septal defect. In the remaining 33 cases (20.5 per cent) the roentgenologic appearance of the heart varied and characteristic features were absent. Of these patients, 18 had tetralogy of Fallot, 8 tricuspid atresia or stenosis, 2 transposition of the great vessels, 2 pulmonic stenosis and single ventricle, 1 pulmonic stenosis and ostium atrioventriculare commune, 1 pulmonic stenosis and patent foramen ovale, and in 1 case the diagnosis was not elucidated.

Postoperatively the roentgenologic appearance of the heart was unaltered in 106 of the 115 cases (92 per cent) in which such assessment could be made. In 6 cases the size of the heart was clearly increased, and in 3 cases the shape of the heart was considerably altered.

The aorta was left-sided in 109 cases (68 per cent) and right-sided in 52 cases (32 per cent). Of the cases with dextropositioned aorta, 49 had tetralogy of Fallot, 1 had tricuspid atresia, 1 pulmonic stenosis and single

ventricle, and 1 pulmonic stenosis and ostium atrioventriculare commune. ABBOTT (1936) and TAUSSIG (1947) found right-sided aorta in about 20 per cent of their cases of Fallot's tetralogy, and this was later confirmed by EEK (1949) and KJELLBERG, MANNHEIMER, RUDHE & JONSSON (1959).

Cardiac volume: In order to determine if the volume of the heart had increased during the postoperative observation period, comparisons were made in the 112 cases in which this volume was determined both before and after shunt operation, irrespective of diagnosis. The relative volume of the heart increased by a mean of 72.1 ± 6.8 ml. Confidence intervals on the 99 per cent level were found at 54 and 90 ml. When the values were corrected for the cases in which death occurred during the follow-up period, the increase was still more significant and the confidence intervals occurred at 60 and 93 ml.

The possibility was considered that the children who died at operation might have differed from the other children as regards preoperative relative cardiac volume. These two groups were therefore compared in this respect. No significant difference was found between the means of the respective groups. Differences between individuals could not be ruled out, however, as the comparison was between different children and not between two values in the same children.

The increase in relative cardiac volume was compared in younger (< 5 years) and in older (≥ 5 years) children. These age groups did not differ significantly, even when variations in observation time were levelled out by subdividing the age groups according to length of follow-up.

Table 16 shows the mean relative cardiac volume before and after operation distributed in 6 age groups. Significant increase thus occurred postoperatively in the age groups 4–7, 9–12 and ≥ 12 years, whereas the increase in the 7–9 year group was not significant. The number of patients (variables) was smallest in this last group, however, and consequently the spread of values was relatively great, which may have tended to confuse differences between mean values.

In table 16 are also shown corresponding mean values in healthy children as reported by CARLGREN & EEK.

Angiocardiography

Preoperatively angiocardiography was carried out in 147 children, venous angiocardiography in 122 cases and selective angiocardiography in 25 cases. For obvious reasons, the studies in the early years were

TABLE 16
Mean Cardiac Volume¹ Before and After Operation and Normal Values

	Age in years						
	1—2	2—4	4—7	7—9	9—12	≥ 12	(12—20)
<i>Preoperative</i>							
Heart volume	(305.7)	299.8	305.5	314.2	340.9	334.0	
No. of cases	7	30	59	24	29	10	
<i>Postoperative</i>							
Heart volume	—	(390.0)	364.1	340.9	381.6	414.8	
No. of cases	—	3	17	16	28	47	
<i>Standard error of means</i>							
Preoperative		± 11.0	± 8.0	± 18.9	± 8.4	± 17.1	
Postoperative		—	± 26.5	± 22.2	± 15.9	± 9.7	
Mean increase		(90.2)	58.6	26.7	40.7	80.8	
<i>Normal values</i> ²							
Heart volume	295	304	310	324	348	369	398
Standard error of mean	± 30.4	± 41.5	± 36.2	± 28.6	± 33.6	± 53.8	± 61.9

¹ ml per m² of body surface.

² according to CARLGREN & EEK (1949). The age groups in this normal series correspond to those in my cases, except that the last 2 groups of normal subjects were, respectively, 12—14 and 14—16 years old.

unsatisfactory by present-day standards. Initially a single projection plane was used and later two right-angled planes in oblique projection. Later still, angiocardiograms were taken simultaneously in frontal and lateral projections. Preoperative and postoperative selective angiocardiography has been done since about 1952 at the various paediatric clinics in which the cases were investigated. The contrast medium was injected *via* a heart catheter under positive pressure *ad modum* Gidlund. The technique otherwise was as described by KJELLBERG, MANNHEIMER, RUDHE & JONSSON in "Diagnosis of Congenital Heart Disease".

Postoperatively 63 selective angiocardiographies and a single venous angiocardiography were performed. In consideration of the risks entailed by angiocardiography, and because it was contraindicated in some cases, the number of such procedures was less than the number of heart catheterizations in the postoperative period. No other form of case selection for angiocardiography was employed. Diagnoses made preoperatively with

inferior technique were frequently revised at the later examinations (table 4). In most cases of tetralogy of Fallot the catheter was placed in the right ventricle for injection of contrast medium. In tricuspid atresia and in a few other cases the catheter was guided into one of the atria.

With the aid of angiocardiology 2 cases of mirror image dextrocardia with tetralogy of Fallot were diagnosed. In 14 cases there was a persistent left superior vena cava, which emptied into the right atrium in 13 cases and into the left atrium in the other case. When contrast medium was injected into the right ventricle, the pulmonary artery and the aorta were simultaneously filled out in the cases of Fallot's tetralogy. The distribution

TABLE 17
Types of Pulmonic Stenosis Distributed According to Diagnosis¹

Diagnosis	No. of cases	Type of stenosis					
		None	Presence uncertain	Valvular	Infundibular	Valvular + infundibular	Atresia
Tetralogy of Fallot	143		25	9	48 (3)	58 (13)	3
Pulmonic stenosis + atrial septal defect	2			2 (1)			
Pulmonic stenosis + ostium atrioventriculare commune	1				1		
Pulmonic stenosis + single ventricle	2			2			
Tricuspid atresia or stenosis	8	3	2	1 (1)		1	1
Transposition of the great vessels	2		1	1 (1)			
Diagnosis obscure	1		1				
Total cases	159	3	29	15	49	59	4

¹ Confirmed by angiocardiology or autopsy.

() = type of stenosis doubtful.

of various types of pulmonic stenosis is shown in table 17. In combined valvular and infundibular stenosis a third ventricle was found in 29 cases and also in 5 cases with only infundibular stenosis. When tricuspid atresia or stenosis was present, it was very difficult to demonstrate possible pulmonic stenosis by means of angiocardiology. In 3 of these cases autopsy showed that no pulmonic stenosis was present, but instead there was a

small ventricular septum defect with partial obstruction of the flow to the right ventricle. Angiocardiography further demonstrated 8 cases of peripheral pulmonic stenosis, 1 of pulmonary artery hypoplasia and 1 of post-operative thrombosis of the left pulmonary artery (fig. 16 and 19). Over-riding of the aorta in the tetralogy of Fallot as seen at angiocardiography in lateral projection may be summarized as follows.

	<i>Degree of over-riding</i>					<i>Total</i>
	<i>Undetermined</i>	<i>None</i>	<i>25 %</i>	<i>50 %</i>	<i>>50 %</i>	
No. of cases	66	11	10	28	29	144

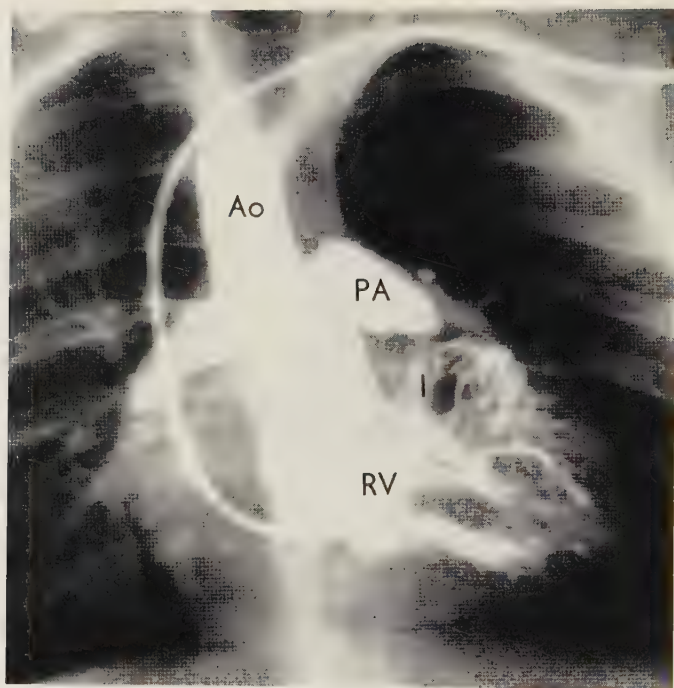
The function of the artificial anastomosis could in many cases be assessed with the aid of angiocardiography in the Blalock as well as in the Potts anastomosis (indirectly in the latter cases). In 7 cases angiocardiography revealed strong evidence of thrombosis in the anastomosis. Patency was demonstrated by passage of contrast medium through the anastomosis in 37 cases. In 6 cases there were indirect signs of function—filling of pulmonary vessels *via* the anastomosis or thinning of contrast in the pulmonary artery at the level of the anastomosis. In 14 cases the function of the anastomosis could not be reliably judged by angiocardiography. Figures 13 to 23 present examples of various types of artificial anastomosis. Figures 13 to 17 show different sizes of functioning right and left subclavian anastomosis. A thrombosed anastomosis with a richly developed net of collateral arteries around it is seen in figure 18. Thrombosis of a left-sided anastomosis and partial thrombosis of the left pulmonary artery together with profuse collateral channels in the left lung are shown in films *a* and *b* of figure 19. In figure 17 the anastomosed subclavian artery is partially stenosed and there is prestenotic dilatation.

The postoperative angiocardiograms demonstrated that the augmentation of the pulmonary circulation was small when the anastomosis was stenotic and also when the subclavian artery that was joined to the pulmonary artery was long and fine-calibred. In such cases the risk of thrombosis is constantly present. In patients with satisfactorily functioning anastomosis the pulmonary vascular bed clearly was better developed than before the operation. A tendency to selective or better ipsilateral contrast filling of the pulmonary vessels was observed in all degrees of functioning anastomosis.

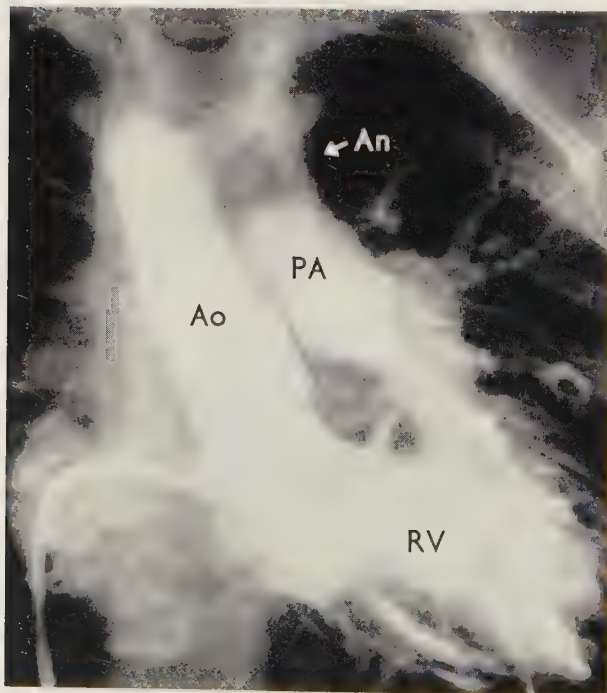
ILLUSTRATIONS

Abbreviations in angiocardiograms

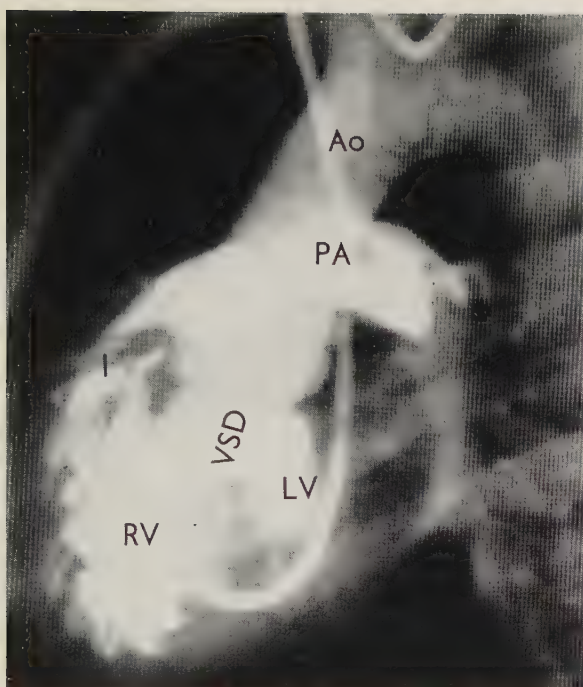
An	=	anastomosis
Ao	=	aorta
Col	=	collateral
I	=	infundibulum
LA	=	left atrium
LV	=	left ventricle
LPA	=	left pulmonary artery
PA	=	pulmonary artery
PV	=	pulmonary valves
RA	=	right atrium
RV	=	right ventricle
RPA	=	right pulmonary artery
VSD	=	ventricular septal defect



"



"



b

Fig. 13. K.-E. D. 3-year-old boy with tetralogy of Fallot. Anastomosis was made between the left subclavian artery and the left pulmonary artery at age 2 years. He was much improved postoperatively and the anastomosis appeared to be patent. Frontal and lateral views *a* and *b* show preoperative angiocardiography from the right ventricle. At postoperative angiocardiography (*c*, frontal view), the right ventricle is richly trabeculated. Severe infundibular and valvular pulmonic stenosis are present. The aorta is wide and shows 50 per cent over-riding. The left-sided anastomosis is seen and there is better contrast filling of the left pulmonary vascular bed.

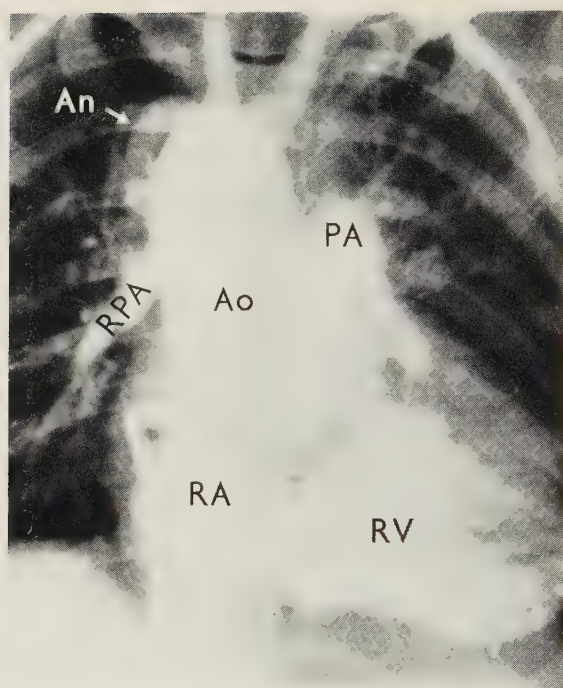


Fig. 14. G. L. 7-year-old girl with tetralogy of Fallot. Anastomosis of the right subclavian to the right pulmonary artery was performed at age 3 8/12 years. Operation was followed by clinical improvement and the anastomosis seemed to function.

Angiocardiographic follow-up with injection of contrast medium into the right ventricle and later into the right atrium shows the ventricle to be enlarged and richly trabeculated. There is infundibular and valvular pulmonic stenosis and also a small third ventricle. The aorta is very wide. The frontal view shows the right-sided subclavian anastomosis.

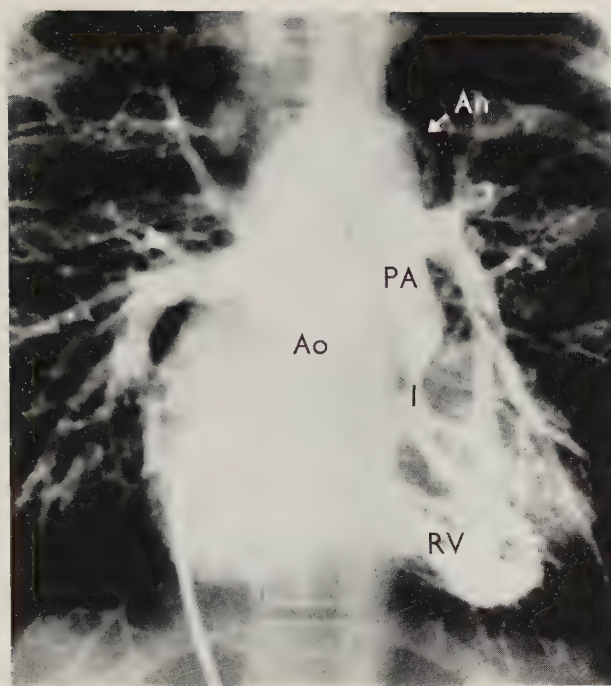
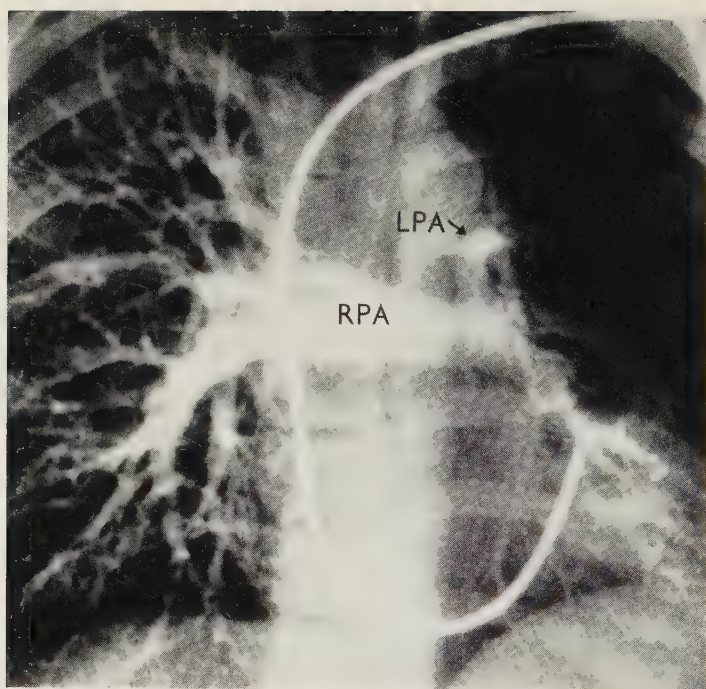


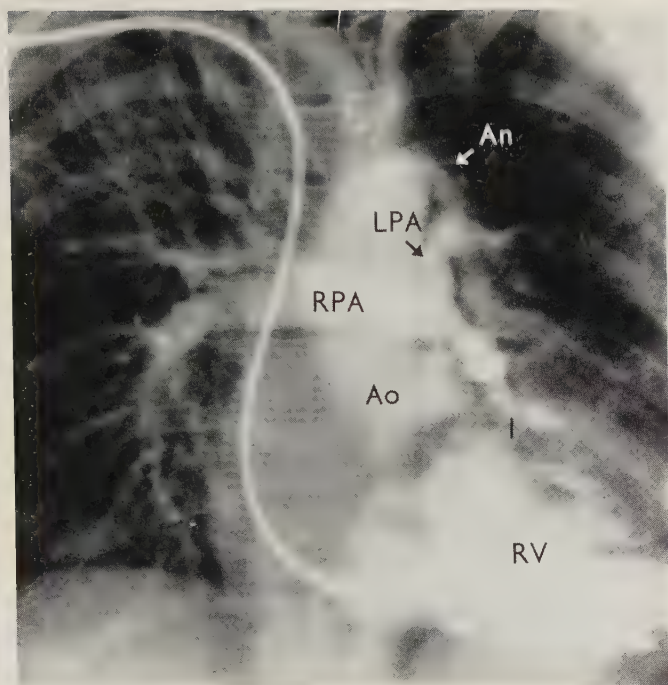
Fig. 15. K. R. 8-year-old girl with tetralogy of Fallot. Anastomosis between the left subclavian and the left pulmonary artery was performed at age 4 10/12 years. Definite clinical improvement followed the operation. Follow-up angiocardiology from the right ventricle shows infundibular and valvular pulmonic stenosis. The aorta is simultaneously filled out. It is dilated and shows about 50 per cent over-riding. The frontal view shows the fine-calibred, left-sided anastomosis and thin pulmonary blood vessels.



a

Fig. 16. M. M. 9-year-old girl with tetralogy of Fallot. Anastomosis between the left subclavian and the left pulmonary artery was performed at age 7 5/12 years. Postoperatively there was distinct improvement.

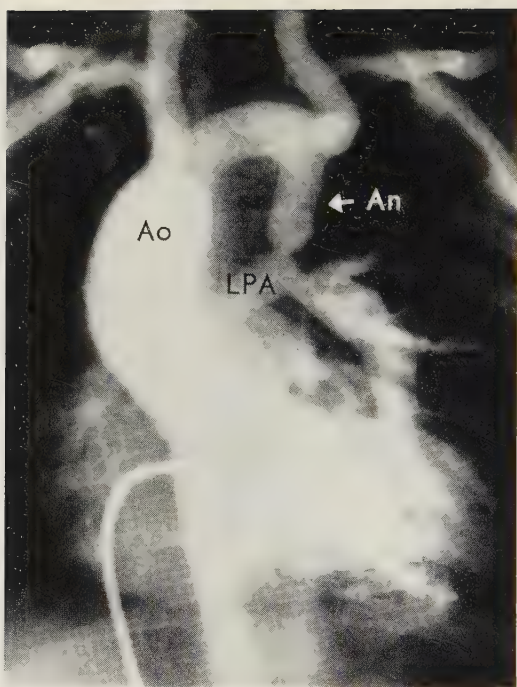
a. Preoperative angiocardiology from the right ventricle shows severe infundibular and valvular stenosis and a small interjacent third ventricle. The main stem of the pulmonary



b

artery is narrow. The right pulmonary artery is wide and the left is hypoplastic. The vascular markings in the left lung are slender and scanty.

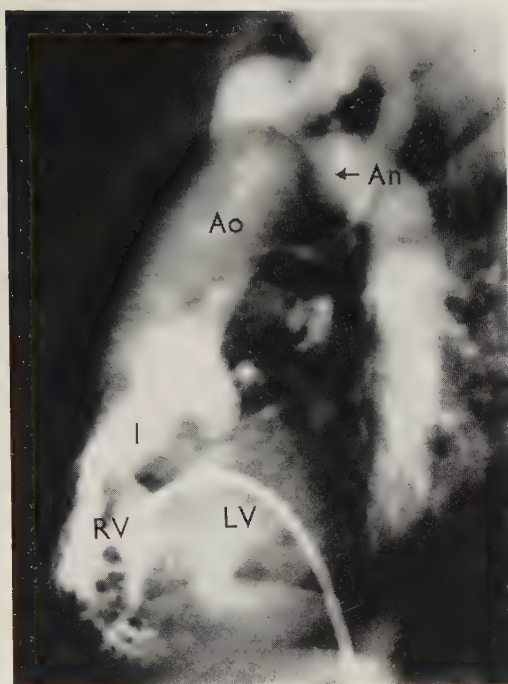
b. Frontal view of functioning artificial anastomosis on the left side. The left pulmonary artery is now wider.



a

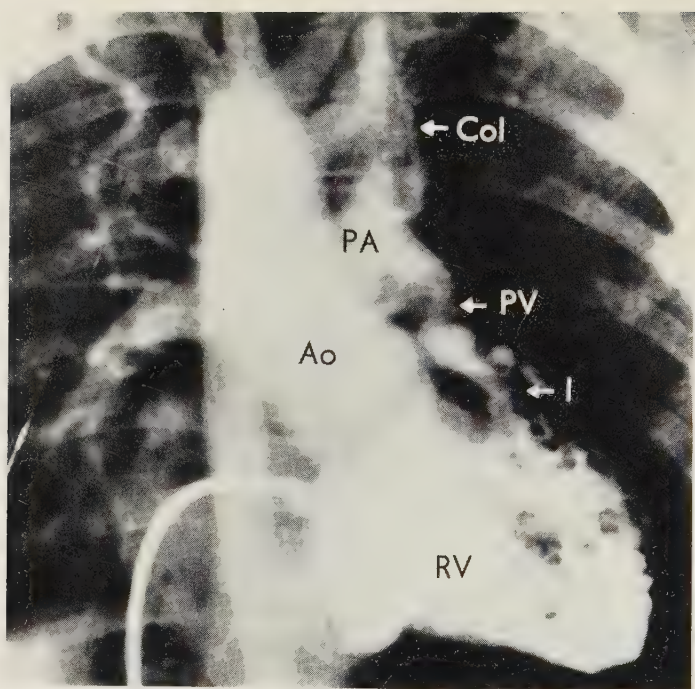
Fig. 17. R. L. 3-year-old boy with tetralogy of Fallot. The left subclavian artery was anastomosed to the left pulmonary artery at age 8 months. Although the child's condition has improved, he is still disabled.

Follow-up angiocardiography from the right ventricle shows the ventricle to be richly



b

trabeculated. Infundibular pulmonic stenosis is present and there is about 50 per cent aortic over-riding. The anastomosis is functioning, but stenosis at the suture site and prestenotic dilatation are present (*a* = frontal view, *b* = lateral view).



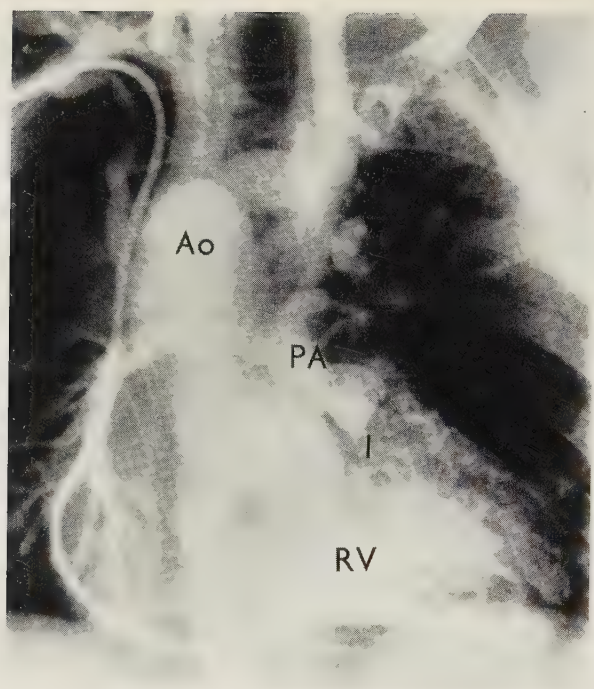
a

Fig. 18. L. B. 6-year-old boy with tetralogy of Fallot. Anastomosis between the left subclavian artery and the left pulmonary artery was performed at age 5 2/12 years. His condition improved at first, but deteriorated later and showed no signs of a functioning anastomosis. Follow-up angiocardiology was performed from the right ventricle. The frontal view *a* shows the enlarged and richly trabeculated ventricle. Severe infundibular and valvular pulmo-



b

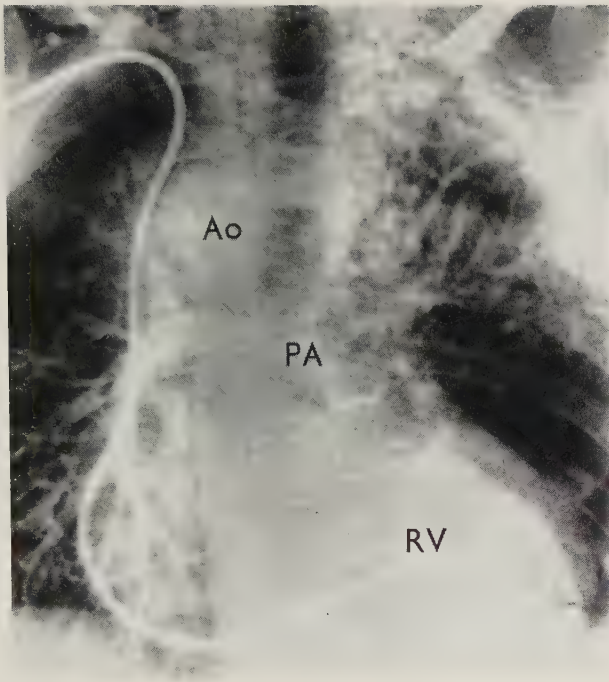
nic stenosis and a small third ventricle are present. The aorta is dilated and 50 per cent over-riding. The anastomosis is blocked, probably by thrombosis. The pulmonary artery shows peg-shaped bulging, and at the site of the anastomosis there are numerous small, irregular vessels. The vascular bed in the upper region of the left lung displays considerable irregularity, indicating collateral circulation. *b*. Close-up of the site of anastomosis.



a

Fig. 19. A. K. S. 12-year-old girl with tetralogy of Fallot. Left subclavian-pulmonary artery anastomosis was done at age 3 9/12 years. No improvement resulted and there were no signs of patency of the anastomosis.

Follow-up angiocardiology was made from the right ventricle. The frontal view *a* shows the ventricle to be richly trabeculated and hypertrophic. Severe infundibular and valvular



b

pulmonic stenosis are seen and also a third ventricle. The aorta, which is over-riding, is filled simultaneously *via* the ventricular septal defect. The anastomosis is thrombosed and is surrounded by a rich net of collateral channels. The left main branch of the pulmonary artery is partially thrombosed. View *b*, taken 2/3 second later, shows enormous collateral circulation to the left lung.

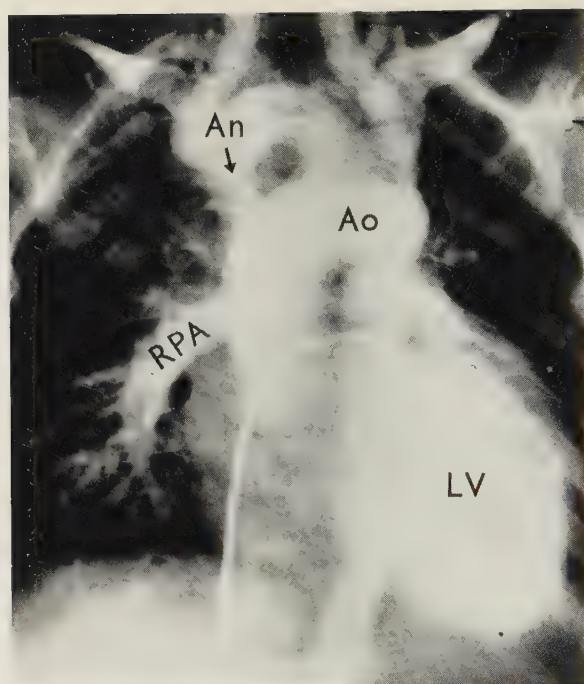


Fig. 20. P. A. W. 9-year-old boy with tricuspid atresia, interatrial and interventricular septal defect and probably also pulmonic stenosis or atresia. At age 3 7/12 years the right subclavian artery was anastomosed to the right pulmonary artery. Postoperatively there was some clinical improvement, but the child is still greatly disabled. Signs of functioning anastomosis are present. Angiocardiographic follow-up was done from the left atrium. The frontal view shows the dilated aorta and the right-sided anastomosis, which is slightly constricted at the pulmonary artery. Both pulmonary arteries are contrast-filled, the right being somewhat wider than the left. The main stem is not visualized.

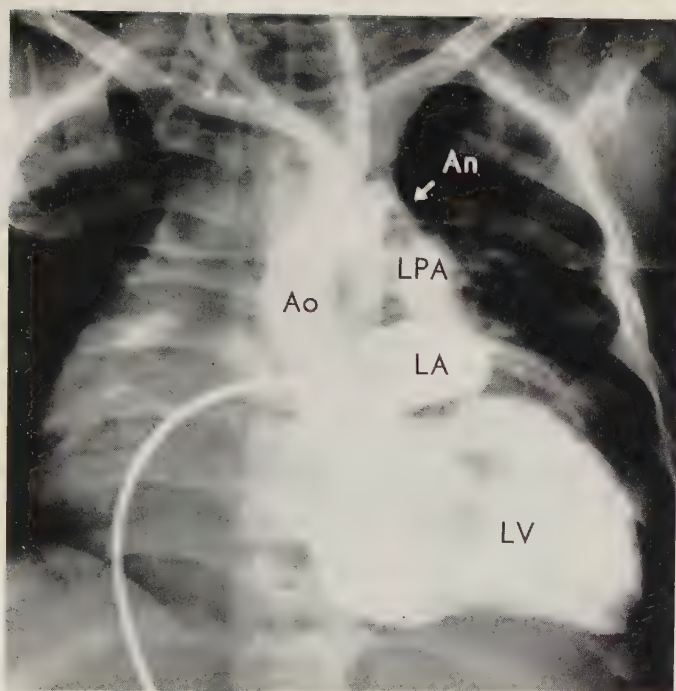
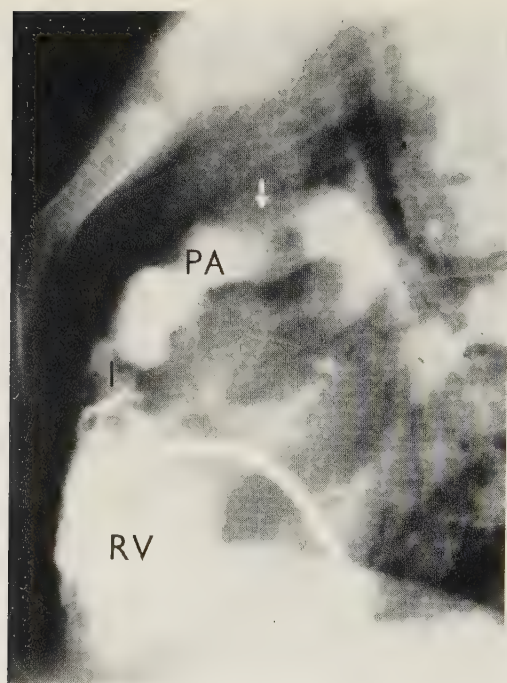


Fig. 21. A. N. 6-year-old girl with relative tricuspid stenosis and insufficiency combined with patent foramen ovale and infundibular and valvular pulmonic stenosis. She died during revision of the first operation. During life the haemodynamic picture suggested tricuspid stenosis or atresia and atrial septal defect with right-left shunt and greatly diminished pulmonary circulation. At age $1\frac{3}{12}$ years left subclavian-pulmonary artery anastomosis was carried out. At first her condition improved, but at the time of follow-up it was poor.

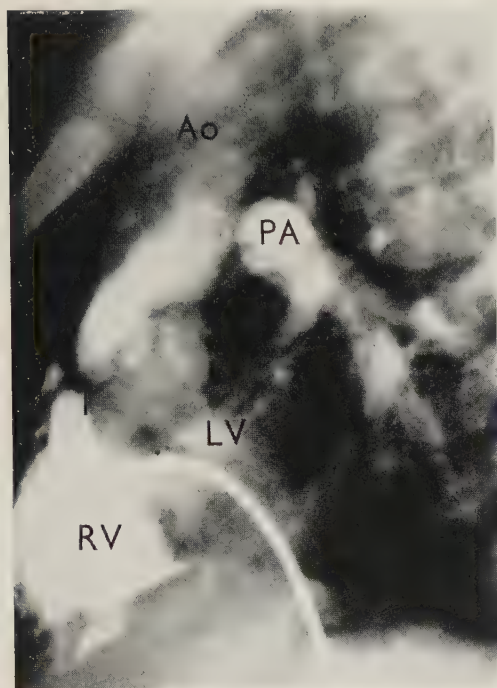
Postoperative angiocardiography was done from the left atrium. On the frontal view the narrow, patent anastomosis is filled from the aorta. Contrast medium reaches the left pulmonary artery from the anastomosis. The right side of the heart, which is not contrast-filled, is greatly dilated.



a

Fig. 22. G. B. 4-year-old girl with tetralogy of Fallot. Direct Potts anastomosis between the aorta and the left pulmonary artery was performed at age 3 1/2 years. Postoperatively her condition was decidedly improved and the anastomosis appeared to function well.

Postoperative angiocardiography shows the right ventricle to be richly trabeculated and enlarged. Severe infundibular and valvular stenosis is present and also a third ventricle.



b

Between the lateral views *a* and *b* there is a time interval of one-third of a second. At the level of the anastomosis the contrast medium in the pulmonary artery is diluted by the blood flow from the aorta (*a*). There is about 50 per cent over-riding of the aorta. Contrast medium passes from the right to the left ventricle *via* the septal defect.

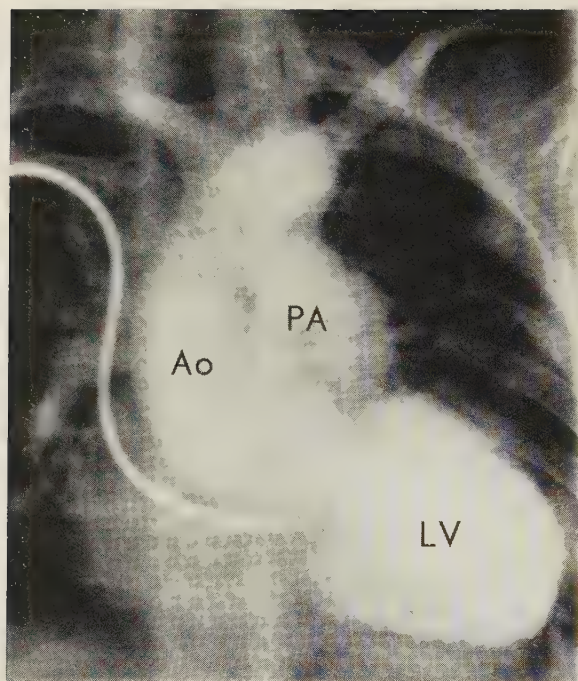


Fig. 23. M. J. 5-year-old girl with tetralogy of Fallot. Operation according to Potts (direct anastomosis between aorta and pulmonary artery) was performed at age 4 1/12 years. The clinical result was good and the anastomosis seemed to function well. Follow-up angiocardiology was done from the left atrium. The frontal view shows the pulmonary artery contrast-filled from the aorta *via* the anastomosis. There is weak contrast filling of the right ventricle *via* the ventricular septal defect.

CHAPTER 9

LATE RESULTS

In direct association with shunt operations there were 22 deaths (13.7 per cent) among the 161 cases. During the observation period, up to the end of 1958, 9 more deaths occurred. The interval between shunt operation and death in these 9 cases ranged from 6 weeks to 7 years and the mean postoperative survival time was 3 6/12 years. The diagnosis was tetralogy of Fallot in 5 cases, tricuspid atresia in 3 cases and pulmonic stenosis with ostium atrioventriculare commune in 1 case. In only 1 of the 9 cases was it considered probable that death resulted from the anastomosis operation and consequent primary heart failure: Autopsy at a provincial hospital showed tricuspid atresia with great enlargement and dilatation of the atria and the left ventricle. The left ventricle communicated with the insignificant and underdeveloped right ventricle *via* a very small septal defect. In 2 cases of tetralogy of Fallot the respective causes of death were meningitis and encephalitis, which were unconnected with the primary cardiac anomalies. Both patients had previously shown some improvement after shunt operation, although the result was not regarded as fully satisfactory. Another patient with tricuspid atresia showed an excellent result from the anastomosis operation but died of brain abscess 6 years later. Pulmonary tuberculosis was the cause of death in a patient with tetralogy of Fallot and probably poor function of the anastomosis. A further patient with tetralogy of Fallot died before a planned revision of the operation could be carried out: Autopsy revealed extensive thromboembolism in the lungs, and the anastomosis too was thrombosed. A patient with Fallot's tetralogy and another with tricuspid atresia died at home, and in neither case was a physician's diagnosis obtained or autopsy performed. A patient with pulmonic stenosis and ostium atrioventriculare commune, in whom no clinical signs of patency of the artificial anastomosis were found at follow-up examination, died later at a provincial hospital: At autopsy thrombosis was observed in the anastomosis and also in the whole of the left pulmonary artery.

In the surviving cases I have reviewed the results of the shunt operations. In 1952, MANNHEIMER, LANDTMAN & HEDQVIST surveyed the results in 78 of the cases in the present series. I have classed the patients who were alive at follow-up into 4 groups according to results of surgery: 1. excellent result, 2. good result, 3. improved, and 4. unimproved.

By excellent results is meant that the patients were acyanotic at follow-up examination, or showed only a suggestion of cyanosis at rest and no or slight cyanosis during exercise. They were able to attend school or work, and in general do what other children or young adults do, with the exception of the more violent forms of activity. The oxygen saturation of the arterial blood rose to between 80 and 95 per cent. In most instances the erythrocyte count, the amount of available haemoglobin and the haematocrit reading returned to approximately normal values.

A good result was considered to have been obtained when the patients were acyanotic at follow-up examination or showed only slight (+) cyanosis at rest and slight to definite cyanosis during exercise. The exercise tolerance of these patients was good, but was not comparable with that in the first group. As a rule the patients were able to lead a fairly normal life, provided that they avoided strenuous activities. The oxygen saturation of the arterial blood rose to between 75 and 90 per cent. The erythrocyte count, the amount of available haemoglobin and the haematocrit became approximately normal or there was slight polycythaemia. The boundary-line between these two groups was not distinct, but cases in which some negative aspect emerged at the follow-up examination were placed in group 2.

Improved means that the patient's condition was improved by the operation, but cyanosis and polycythaemia of varying degrees persisted. The physical capacity was clearly subnormal, but was better than before the operation. The oxygen saturation of the arterial blood was not as a rule less than 70 per cent.

The group of patients who were judged to be unimproved by operation includes those who derived no benefit from surgery and also those in whom an initial improvement was followed by gradual deterioration to the pre-operative level.

Prior to the final summation of results, I divided up the cases in the 4 groups according to diagnosis, presence or absence of continuous murmur (as a presumptive sign of anastomosis patency) and relative heart volume. The findings then were as follows.

	No. of cases	Mean age (years)	Continuous murmur	Mean heart volume (per m ² body sur- face)
<i>Tetralogy of Fallot</i>				
1. excellent	31 (29%)	13	31	400
2. good	37 (34%)	11	37	390
3. improved	22 (20%)	11	20	350
4. unimproved	18 (17%)	9	5	320
<i>Tricuspid atresia</i>				
2. good	1	5	1	570
3. improved	1	18	1	430
4. unimproved	2	4	1	(370 & 680)
<i>Transposition of great vessels</i>				
3. improved	1	7	1	410
4. unimproved	1	14	1	470
<i>Pulmonic stenosis with patent foramen ovale</i>				
3. improved	1	18	1	520

It is seen that in the tetralogy of Fallot the relative heart volume increased in proportion to the degree of postoperative improvement. Presumably the cardiac volume was also correlated to the size of the shunt (i.e., of the anastomosis). Among the patients with Fallot's tetralogy who were unimproved by shunt operation, only a minority had continuous murmur, and this suggests obstruction or blockage of the anastomosis in most cases. In the other represented conditions the small numbers of cases make conclusions difficult. It is probable that the relative heart volume at follow-up examination was higher in these cases than in the patients with tetralogy of Fallot.

Additional to the patients in this survey were those who lived abroad and could not present themselves for postoperative examination, but were asked by letter to report their state of health. From the detailed replies that were received, it seemed that of 13 cases of morbus caeruleus in which shunt operations were performed, 5 could be classed as having an excellent result (group 1), while 5 had a good result (group 2) and 3 were improved (group 3).

Concerning 2 other foreign patients, no information at all was available. Although on discharge from hospital in Sweden these patients were

definitely improved, it was not possible to classify them according to results of operation.

After the approximation of adding the written replies to the clinical findings, the following results were obtained in the cases in which the postoperative condition was known.

	<i>Patients alive at follow-up</i>	<i>Patients alive at follow-up + late deaths</i>
Excellent or good	79 (62%)	79 (58%)
Improved	28 (22%)	28 (20%)
Unimproved	21 (16%)	21 (15%)
Dead		9 (7%)
Total	128	137

Twenty of the patients who were improved or unimproved by the initial operation underwent revision of the anastomosis during the follow-up period. The results of these later operations are not included in the follow-up of the initial shunt operations.

CHAPTER 10

STATISTICAL CALCULATIONS

In the statistical analyses the following formulas and tests were used.

1. For calculation of mean values:

$$m = \frac{\sum x_i}{n} \text{ where } \sum x_i \text{ is the sum of values and } n = \text{the number of terms.}$$

For calculation of the spread of a variable:

$$S^2 = \frac{1}{n-1} \sum_i (x_i - m)^2 \quad (S = \text{the standard deviation of the variable})$$

$$= \sqrt{\frac{1}{n-1} \sum_i (x_i - m)^2}$$

For calculation of the standard deviation of the mean:

$$\Sigma(m) = \frac{S}{\sqrt{n}}$$

For calculation of the spread of the difference between 2 variables (mean values):

$$S_p^2 = \frac{(n_1 - 1) S_1^2 + (n_2 - 1) S_2^2}{n_1 + n_2 - 2} \text{ where } n_1 = \text{the number of terms for one variable and } n_2 = \text{the number of terms for the other variable.}$$

S_1^2 and S_2^2 are the corresponding spreads:

$$S_1^2 = \frac{1}{n_1 - 1} \sum_i (x_i - m_1)^2 \text{ and } S_2^2 = \frac{1}{n_2 - 1} \sum_i (y_i - m_2)^2$$

m_1 and m_2 are the means of the respective variables:

$$m_1 = \frac{\sum x_i}{n} \quad m_2 = \frac{\sum y_i}{n}$$

2. To test significance of difference between 2 means:

a) A preoperative and a postoperative value from the same case are compared.

The following quantity is calculated:

$$t = \frac{m_1 - m_2}{S_q / \sqrt{n}} \text{ where } S_q = \sqrt{\frac{1}{n-1} \sum_i (d_i - \bar{d})^2}$$

and d_i = the difference between the preoperative and postoperative value for each individual. $\bar{d} = \frac{\sum_i d_i}{n}$

If t exceeds the tabulated t value for $n-1$ degrees of freedom on 95 per cent level, significance is present. A difference is called clearly significant when the t -value has exceeded the 99 per cent level in the table.

- b) Two categories of individuals are compared before and after operation.

The following quantity is calculated:

$$t = \frac{m_1 - m_2}{S_p \sqrt{\frac{1}{n_1} + \frac{1}{n_2}}} \text{ where } S_p = \sqrt{\frac{(n_1 - 1) S_1^2 + (n_2 - 1) S_2^2}{n_1 + n_2 - 2}}$$

If t exceeds the tabulated t -value for $n_1 + n_2 - 2$ degrees of freedom on 95 per cent level, significance is present. When the 99 per cent level has been exceeded the difference is called clearly significant.

(If t is negative, the same applies, with reversed signs).

3. To test differences between values before and after operation, divided into 2 categories:

The following quantity is calculated:

$$X^2 = \frac{(n_1 + n_2) n_1 n_2}{(d_1 + d_2) (n_1 + n_2 - d_1 - d_2)} \left(\frac{d_1}{n_1} - \frac{d_2}{n_2} \right)^2$$

where d_1 and $n_1 - d_1$ are the numbers of individuals before operation divided into 2 categories (e.g., age groups) and d_2 and $n_2 - d_2$ are the corresponding numbers after operation.

If X^2 exceeds the tabulated X^2 -value for 1 degree of freedom on 95 per cent level, significance is present. If the 99 per cent level is exceeded the difference is clearly significant.

CHAPTER 11

GENERAL DISCUSSION

It is no exaggeration to say that the classical operations in which an opening is made from a branch of the aorta or from the aorta itself to a pulmonary artery have revolutionized the treatment of cyanotic congenital heart disease such as the tetralogy of Fallot, tricuspid atresia and other similar anomalies in which the pulmonary circulation is diminished, and have given reasonably good health to thousands of patients for whom the outlook otherwise was only progressive disability and early death. In recent years, open-heart surgery for complete corrective repair has superseded these shunt operations for the treatment of Fallot's tetralogy in many centres. Because of the relatively high mortality which the radical operations still carry, however, it is necessary to consider whether or not shunt operations, despite their palliative nature, still merit a place in modern cardiac surgery. A number of reports of "late results" and "long-term results" have been published in order to illustrate the advantages and drawbacks of these operations. Most writers now seem to agree that open-heart surgery is the method of choice in cases which fulfil the criteria of suitability. In other cases the indications for artificial anastomosis remain as before.

The primary surgical mortality in shunt operations has shown wide variations in different case series and has also been reported in different ways. TAUSSIG & BAUERSFELD (1953) stated that the surgical mortality was 15.7 per cent in the first 1,000 cases of pulmonic stenosis or atresia in which anastomosis was made between the systemic and the pulmonary circulation. CAMPBELL & DEUCHAR (1953) reported 12 per cent primary operative mortality in 200 cases of morbus caeruleus. The corresponding figure found by NADAS (1957) was 11 per cent. According to DONZELOT ET AL. (1957), 16.2 per cent of 351 patients died in association with shunt operations. POTTS ET AL. (1955) wrote that in the first 100 cases of aortico-pulmonary anastomosis for the tetralogy of Fallot the surgical mortality was 9 per cent. SHUMACKER & MANDELBAUM (1960) reported the remarkably

low mortality rate of 4.7 per cent in a series of tetralogy of Fallot, in which 86 patients underwent subclavian-pulmonary artery anastomosis and 29 aortico-pulmonary artery anastomosis.

Of my 161 patients, 13.7 per cent died in direct association with shunt operation. As in many other series, the surgical mortality was highest in the first years of the study period, but fell as the technique was stabilized. In the patients who were less than 5 years old at operation, the surgical mortality was higher than in the older patients. A similar tendency was reported by other writers. POTTS, for instance, stated that 17.1 per cent of the children younger than 3 years died, as compared with 4.6 per cent of the older patients. CAMPBELL & DEUCHAR reported that the surgical mortality in children younger than 4 years was 17 per cent and that the rate then fell until it reached 5 per cent in patients aged 15 to 19 years. According to GROSS (1953), the mortality in children less than 3 years old was 30 per cent, whereas in older children it was 7.1 per cent.

Late results are difficult to compare in published series of cases, since the criteria are exceedingly varied, as is the period of observation. TAUSSIG & BAUERSFELD reported that of the 1,000 patients who were operated on between 1944 and 1950, the results were good in 773 and fair in 39, while 31 were unimproved and 157 patients died at or immediately after operation. CAMPBELL & DEUCHAR found that the results of shunt operation were excellent in patients with tetralogy of Fallot. Many of the children had been seriously ill, but only 8 per cent died and 75 per cent benefited greatly. In the smaller number of more complex lesions, such as tricuspid atresia, dextrocardia and pulmonic atresia, the results were much less good: The total mortality was nearly 30 per cent, and only 35 per cent obtained a good result. In continued follow-up, CAMPBELL (1958) noted that, of the patients in whom the result of operation was good, 80 per cent maintained this result for an average period of 7 years after operation. He thought that perhaps half of those with good results might be reasonably well 20 years postoperatively. POTTS ET AL. (1955) reported that of the first 100 cases in which anastomosis was made between the aorta and the pulmonary artery, 68 per cent showed a good result and 16 per cent a fair result 6 to 8 years later. Five per cent improved after operation but died later.

Despite the variety of criteria, the late results in the literature seem to be in fairly good agreement with those in my account of late results, when the observation period is taken into account. There appears to be unanimity of opinion that the best results of shunt operations are obtained in the tetralogy of Fallot. The results in tricuspid atresia must be regarded as distinctly poorer, although the comparative rarity of the condition makes

the reported series relatively small. With respect to both physical capacity and survival time, tricuspid atresia responds less well to shunt operation than does the tetralogy of Fallot. Moreover, when the shunt through the anastomosis is large, the risk of progressive cardiac enlargement and resultant insufficiency is greater because of the strain on the left side of the heart before operation. My cases of other conditions than the tetralogy of Fallot were too few to permit conclusions.

In their follow-up of 100 cases 11 to 13 years after operation, PAUL, MILLER & POTTS (1961) performed heart catheterization in 18 patients postoperatively. They found greatly elevated mean pulmonary artery pressure (45 to 85 mm Hg) and increased pulmonary vascular resistance in 3 cases. The latter was considered to be secondary to a powerful shunt through the artificial anastomosis. Only 13 of my patients underwent the Potts operation. In 5 of them postoperative heart catheterization was performed. Only 1 showed a tendency to pulmonary hypertension (mean pressure 20 to 25 mm Hg). The relative heart volume and the electrocardiogram in this patient were unchanged after operation and showed no signs of progressive cardiac enlargement.

Seventeen patients with late haemodynamic complications of anastomotic surgery for the tetralogy of Fallot were studied by ROSS, TAUSSIG & EVANS (1958). According to these writers, two syndromes may develop as late haemodynamic complications of such operations. Both can be related to a shunt of greater than ideal size. The first is characterized by cardiac enlargement and congestive failure similar to that seen in patients with arteriovenous fistulae. The second is characterized by pulmonary hypertension, aneurysmal dilatation of the pulmonary artery, decreasing shunt, return of cyanosis, cardiac failure and death. In my patients there was significant increase of the relative heart volume after operation. Prior to operation the heart volume as a rule was less than the normal for the relevant age. After operation the values rose to slightly higher than normal. The increase in relative heart volume seemed to be in proportion to the degree of general improvement. The groups in which the results were classed as excellent or good had the largest shunt through the anastomosis. In the cases classed as improved or unimproved, the heart volume was less and the shunt was smaller or nonexistent. The 4 cases in which I found postoperative pulmonary artery hypertension belonged to the group with the greatest degree of postoperative improvement. In 2 of these patients (Cases 102 and 104) the shunt was definitely excessive. In both cases the relative heart volume was considerably greater than the normal for the age. At autopsy Case 102 was shown to have tricuspid stenosis, pulmonic

stenosis and patent foramen ovale. The artificial anastomosis had increased the work of the heart so that insufficiency occurred. Excessive width of anastomosis between the carotid artery and the pulmonary artery in Case 139 led to progressive cardiac enlargement with attacks of paroxysmal tachycardia. Cases 102 and 139 died after the end of the observation period. No other cases of the syndromes described by ROSS, TAUSSIG & EVANS occurred in my series of cases.

In the patients with tetralogy of Fallot, the action potential of the left ventricle increased significantly after operation. This increase, like that of the relative heart volume, was of moderate degree. PAUL, MILLER & POTTS found electrocardiographic signs of combined ventricular hypertrophy in two-thirds of their patients postoperatively, indicating that there had been considerable increase in left ventricular work following the surgical anastomosis with its attendant increase in pulmonary blood flow. In POTTS' original case series the anastomoses seem to have been made considerably wider than those in my patients. This presumably explains the absence of left ventricular hypertrophy in the postoperative electrocardiograms of my patients.

That the size of the heart increases fairly soon after shunt operations and becomes stabilized after a year or two was demonstrated by a number of writers (BLALOCK & TAUSSIG 1945, MANNHEIMER 1949, CAMPBELL & DEUCHAR 1953). Thereafter it remains relatively unchanged during 8 to 10 years (CAMPBELL 1958). The risk of progressive cardiac enlargement increases with the size of the shunt. Whether or not the anastomosis may grow with the child is a question that cannot be conclusively answered. The results of my follow-up angiocardigraphic studies indicate a positive answer. Contradictory, however, are the poorer clinical results in the children younger than 5 years at operation as compared with the older children. One may nevertheless assume that a relatively wide anastomosis is more likely to provide an adequate shunt later in life than is a narrow anastomosis with small diversion of blood to the pulmonary circuit.

Although nowadays open-heart surgery with resection of pulmonic stenosis and closure of interventricular septal defect is generally advocated in the tetralogy of Fallot, there are still cases in which establishment of a shunt between the systemic and the pulmonary circulation can be recommended. A Blalock or Potts operation may be advisable for children who are considered to be too young for radical correction and who are in too poor a condition to await a more suitable age for such surgery. When there is much over-riding of the aorta or an underdeveloped left ventricle, or in cases with hypoplasia of the pulmonary artery, an anastomotic pro-

cedure may be preferable. For a severely ill and cyanosed child, early establishment of an artificial shunt may be necessary to overcome a critical situation. If the shunt operation can be delayed until the child is older than eight years, however, the attendant risks will be less and there will be a better prospect of satisfactory function. If one intends to perform a radical operation when the child's condition has been improved by the anastomosis, a subclavian-pulmonary artery communication is better than a Potts anastomosis, which is technically more difficult to close. After the cardiopulmonary circulations have been balanced up for a few years, definite correction may become feasible. According to KIRKLIN, PAYNE & DU SHANE (1960) the probability of surviving open cardiac repair is high for patients with previously constructed anastomotic operations.

Tricuspid atresia with reduced pulmonary circulation has not hitherto been successfully corrected by intracardiac operation. In consequence, shunt operations are recommended, although the results generally are poorer than in the tetralogy of Fallot. Transposition of the great vessels with diminution of the pulmonary circulation or truncus arteriosus communis are sometimes considered suitable for artificial anastomosis, but the long-term results are seldom good. The trilogy of Fallot (pulmonic stenosis with interatrial septal defect or patent foramen ovale) should preferably be treated by open repair. One such patient in my series (Case 106) was submitted to surgical anastomosis under the erroneous diagnosis of tetralogy of Fallot and died during a later attempt at surgical correction.

GENERAL SUMMARY

The operation in which an artificial communication is created between the aorta and the pulmonary artery was introduced by BLALOCK and TAUSIG in 1944 and may be regarded as a milestone in cardiac surgery. The operation was designed as a palliative measure to increase the flow of blood to the lungs, which is diminished in the tetralogy of Fallot, tricuspid atresia and haemodynamically similar congenital malformations, and thereby to improve the arterial oxygen saturation in the systemic circulation.

The aim of my investigation was to assess the results of shunt operations in cases of morbus caeruleus. The postoperative condition of the surviving patients was investigated with respect to physical development, exercise tolerance, cardiovascular status and function of the anastomosis.

The series consisted of 161 cases of morbus caeruleus, in all of which the pulmonary circulation was reduced. After special investigations, shunt operations were performed at Kronprinsessan Lovisa's Children's Hospital or at Sabbatsberg Hospital during the period 1947 to 1957. In 148 cases the Blalock-Taussig operation was performed, and in 13 cases direct anastomosis was made between the aorta and the pulmonary artery according to Potts. When possible, the follow-up investigations included heart catheterization and angiocardiography, in order to determine if the shunt operations were followed by secondary hypertension in the pulmonary artery. When progressive cardiac enlargement occurred after the operation, I tried to elucidate the cause. Shunts created during early infancy were studied with respect to functional efficiency later in life, when demands on the circulation had increased.

The postoperative investigations were made by me in 100 cases at Kronprinsessan Lovisa's Children's Hospital. Ten patients were examined at other hospitals in Sweden and 6 were examined at Finnish or Norwegian hospitals. Of the remaining 19 survivors, most of them foreign subjects, all but 2 presented written reports concerning their state of health. Ninety-seven patients were hospitalized for the follow-up investigations, and of these patients 89 were studied by heart catheterization and 63 by selective angiocardiography.

The follow-up studies were concluded at the end of 1958. The observation period from the date of operation ranged from 4 months to 10 10/12 years. The mean observation period was 5 4/12 years and the median 5 10/12 years.

Twenty-two patients (13.7 per cent of the series) died in association with the Blalock-Taussig operation. The surgical mortality was higher in children younger than 5 years as compared with older children. There was no sex preponderance in surgical mortality. The influence of variations in the surgical procedure is discussed. The causes of death are analyzed and also the transient surgical complications and their symptomatology.

There were 9 late deaths during the follow-up period. These cases are discussed with respect to the nature of their congenital malformations, causes of death and duration of survival.

Slightly more than 12 per cent of the children were prematurely born. This is approximately double the incidence of prematurity in a general Swedish population.

The maternal age and the order among siblings did not seem to be significant for the congenital cardiac anomalies. Nor was there any striking familial incidence of such anomalies.

One of the most prominent symptoms of morbus caeruleus is cyanosis. Four grades of cyanosis were recognized. In 64 per cent of the children cyanosis was manifest before the age of 7 months. As a rule it was the first sign of cardiac disorder and appeared before the diagnosis was medically established. After shunt operation the severity and incidence of cyanosis were greatly diminished. The best results as regards cyanosis were obtained in children who were older than 8 years at operation. The physical capacity as reported by patients and relatives, and the walking capacity as judged by me, showed considerable improvement after shunt operations when age was taken into consideration.

Systolic murmur was heard before operation in 157 of the 161 cases. In 1 of the other cases there was only a diastolic murmur, in 1 a continuous murmur, and in 2 cases no murmur was heard before operation. Phonocardiographic follow-up showed the preoperative murmur to be unchanged in 96 per cent of the examined cases. In the remaining 4 per cent the position of the murmur had shifted from the middle to the first half of systole. In 86 per cent there was an additional continuous murmur, which was interpreted as a sign of functioning anastomosis.

After anastomosis between the subclavian artery and the pulmonary artery, the blood pressure in many cases became measurable also on the side of the operation. This was considered to be a result of the collateral

circulation. The pressure in the pulmonary artery could be measured in 42 cases before operation and in 44 cases after operation. In 16 cases readings were available both before and after operation. The preoperative pressure in the pulmonary artery invariably was normal or subnormal. In the cases in which comparisons could be made, there was a statistically significant rise in systolic and diastolic pressure after operation. The pulse pressure in the pulmonary artery was also increased after operation, but the rise was not statistically significant. Pulmonary hypertension probably is not of primary importance for the prognosis after shunt operation for morbus caeruleus. The oxygen saturation in the systemic circulation increased significantly after operation and the haemoglobin, correlated to the oxygen saturation, showed a tendency to fall towards normal values.

Comparisons were made of preoperative and postoperative electrocardiograms. In statistical analysis of the atrial complex, no significant change in amplitude was found after operation over the right side of the heart, but there was significant diminution over the left side. The ventricular complex likewise was unaltered over the right side of the heart, but the amplitude over the left side was significantly increased. The change over the left ventricle is attributed to the increased work of the ventricle due to augmentation of blood flow, through the shunt, to the left side of the heart. The absence of increase in amplitude over the left atrium is not comparable from the haemodynamic aspect.

The relative heart volume showed after shunt operations a mean increase of 72.1 ± 6.8 ml. Confidence intervals on the 99 per cent level were found at 54 and 90 ml. As regards increase in relative heart volume, there was no difference between younger and older patients, even when length of observation time was taken into consideration. As compared with the values for different age groups in the healthy children studied by CARLGREN & ECK, the relative heart volume in my cases generally was less before operation. After operation the values in my patients were significantly higher than the values for healthy subjects of corresponding age. Moderate increase in relative heart volume was thus demonstrated after shunt operations for morbus caeruleus.

Angiocardiography was performed before operation in 147 cases. At follow-up examination angiocardiography with injection of contrast medium through the heart catheter (selective angiocardiography) could be performed or was regarded as permissible in 63 cases. In 37 of these cases patency of the anastomosis was directly demonstrated. The condition in such cases, and also in the cases in which patency was not demonstrated, is discussed.

The follow-up study thus showed encouraging results from shunt operations. From this it is inferred that shunt operations are justifiable in cases of the tetralogy of Fallot or tricuspid atresia in which for various reasons radical correction is not feasible.

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